Results of Tympanoplasty in Children With Primary Ciliary Dyskinesia

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Objective: To assess the results of tympanoplasty in children with primary ciliary dyskinesia complicated by tympanic perforation or cholesteatoma with hearing loss and/or recurrent otorrhea.

Design: Retrospective study. Postoperative follow-up of 26.3 months in the type 1 tympanoplasty group and 46 months in the child with cholesteatoma.

Setting: Hospitalized care, referral center.

Patients: Seven children with primary ciliary dyskinesia, complicated in 6 children by 9 tympanic perforations (3 bilateral perforations) and in 1 child by an attic cholesteatoma.

Results: After 9 type 1 tympanoplasties, the grafts were intact in 9 ears, with no recurrence of otorrhea, but serous otitis media was present in 6 of the 9 ears. Auditory improvement was significant, with an average gain of 17-dB hearing level in speech frequencies. After a canal wall-down tympanoplasty with mastoidectomy for attic cholesteatoma in 1 ear, the cavity that was operated on showed no signs of otorrhea or residual cholesteatoma after a follow-up of 46 months.

Conclusion: In children with primary ciliary dyskinesia, tympanoplasty has a high probability of graft success and auditory improvement, despite the frequent recurrence of serous otitis media.


PRIMARY CILIARY dyskinesia (PCD) is an autosomal recessive disease that is manifested clinically by chronic rhinosinusitis, bronchiectasis, and occasionally situs inversus, which make up the classic triad of Kartagener syndrome. In 1976, Afzelius and Pedersen and Mygind demonstrated immobility of the cilia and an absence of the dynein arms in the cilia of the respiratory mucosa and the flagella of spermatozoa in patients with Kartagener syndrome. The concept of the immotile cilia syndrome, later called primary ciliary dyskinesia, was introduced by Eliasson et al, and also included less severe ultrastructural abnormalities of cilia that led to ciliary hypomobility and were associated with the same clinical picture. In 1978, Fischer et al demonstrated the existence of ciliary abnormalities in the mucous membrane of the middle ear, identical to those in the bronchial mucous membrane. An associated chronic serous otitis media (SOM) has been reported by many authors in cases of disease involving the sinuses and the bronchial tree. Few authors have taken a specific interest in the pathological features of the middle ear in children. According to a few published series of children younger than 15 years, involvement of the middle ear seems to be almost universal. Jachrdsdorfer et al described a high level of tympanic perforation (4 of 6 children) with recurrent episodes of otorrhea. The aim of our study was to assess the benefits that can be expected from tympanoplasty in children with PCD complicated by tympanic perforation or cholesteatoma with hearing loss and/or recurrent otorrhea.

RESULTS

Before the operation, the patients’ clinical symptoms consisted of recurrent episodes of otorrhea associated with hearing loss in 8 ears and isolated hearing loss in 2 ears. Patients 1, 2, 4, 5, and 6 had undergone an adenoidectomy in early childhood, and 6 of the 9 perforated ears had previously undergone 1 to 3 insertions of ventilating tubes (which had been removed more than 20 months before tympanoplasty in every case).
PATIENTS AND METHODS

From August 1988 to May 1995, 10 tympanoplasties were performed in 7 patients with PCD in the Pediatric Otolaryngology and Head and Neck Surgery Department of Armand-Trousseau Children’s Hospital, Paris, France. Three patients presented with a unilateral tympanic perforation, 3 with a bilateral tympanic perforation, and 1 with a cholesteatoma. Data collected on these 7 patients were reviewed. The variables reviewed were age, sex, medical history, indications for surgery, preoperative otoscopy findings, preoperative audiometric data, type of surgery, postoperative otoscopy symptoms and findings, and postoperative audiometric data.

The age of the patients at surgery ranged from 7 to 13.6 years, with a mean age of 10.3 years. Three patients were boys and 4 were girls. The diagnosis of congenital ciliary dyskinesia was made using electron microscopy in 5 cases, or from highly suggestive clinical findings (Kartagener syndrome) in 2 cases. All patients had a complete ear, nose, and throat examination and audiometric evaluation before the operation. Audiometric evaluation was carried out with a diagnostic audiometer in a soundproof room, with recording of pure-tone thresholds using earphones. Student t-tests were used to compare preoperative and postoperative audiometric data.

Two types of surgical interventions were performed: a type 1 tympanoplasty using an endaural approach, and an underlay graft using the temporal fascia with or without a canal wall-up mastoidectomy; or a canal wall-down tympanoplasty with mastoidectomy using an endaural approach, and a graft of the temporal fascia.

In 9 ears, preoperative otoscopy revealed a tympanic perforation (3 children had unilateral perforation and 3 had bilateral perforation) that was located anteriorly (4 ears), inferiorly (1 ear), or that was subtotal (4 ears) (Table 1). The atrial mucosa was inflamed in 4 ears and normal in 5 ears. In these 9 ears, the last episode of otorrhea dated from at least 2 months previously. In 1 child, a cholesteatoma of the epitympanic recess was present, with otorrhea and an intact tense part of the tympanic membrane. Preoperative and postoperative audiometric data are detailed in Table 2.

In the 9 ears with tympanic perforation, a type 1 tympanoplasty was performed, combined with canal wall-up mastoidectomy in 4 ears. With a mean follow-up of 26.3 months, the tympanic membrane was closed in all 9 ears. None of the patients had postoperative otorrhea, regardless of whether a mastoidectomy was performed. In 3 ears, the tympanic membrane was considered normal with a residual air-bone gap inferior to a 10-dB hearing loss. In 6 ears, SOM was present: otoscopy showed retrorotympanic fluid, the tympanometry curve was flat, and the air-bone gap was superior to a 10-dB hearing loss in 5 of 6 ears (postoperative audiometry was not available in 1 ear). None of these cases developed a retraction pocket during the follow-up period. There was no significant difference between the preoperative and postoperative speech frequency bone conduction, or between the preoperative and postoperative 4-kHz bone conduction. Despite recurrent SOM in 6 of 9 ears, the speech frequency air conduction was significantly improved after the operation, and none of the patients required intubation in the postoperative period.

Patient 7 had an attic cholesteatoma revealed by otorrhea and hearing loss (Table 1). He underwent a canal wall-down tympanoplasty with conservation of an intact but hypomobile stapes (tymanosclerosis). Speech frequency air conduction remained unchanged after the operation (45 dB) while mean speech frequency bone conduction was 8 dB before the operation and 5 dB after the operation. After a follow-up of 46 months, there has not been any postoperative otorrhea or development of a residual cholesteatoma.

Table 1. Case Descriptions*

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Operated Ear</th>
<th>Age at Surgery, y</th>
<th>Symptoms</th>
<th>Otoscopy Findings</th>
<th>Size of the Perforation, mm</th>
<th>Preoperative Air-bone Gap, kHz</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>R</td>
<td>13.6</td>
<td>RO and HL</td>
<td>Anterior perforation</td>
<td>0.25</td>
<td>30</td>
</tr>
<tr>
<td>2</td>
<td>L</td>
<td>10.5</td>
<td>RO and HL</td>
<td>Subtotal perforation</td>
<td>Subtotal</td>
<td>28</td>
</tr>
<tr>
<td>3</td>
<td>R</td>
<td>11.3</td>
<td>RO and HL</td>
<td>Subtotal perforation</td>
<td>Subtotal</td>
<td>29</td>
</tr>
<tr>
<td>4</td>
<td>L</td>
<td>7.3</td>
<td>RO and HL</td>
<td>Inferior perforation</td>
<td>0.5</td>
<td>31</td>
</tr>
<tr>
<td>5</td>
<td>R</td>
<td>8.3</td>
<td>RO and HL</td>
<td>Subtotal perforation</td>
<td>Subtotal</td>
<td>25</td>
</tr>
<tr>
<td>6</td>
<td>R</td>
<td>12.5</td>
<td>RO and HL</td>
<td>Inferior perforation</td>
<td>0.25</td>
<td>45</td>
</tr>
<tr>
<td>7</td>
<td>L</td>
<td>10.6</td>
<td>RO and HL</td>
<td>Inferior perforation</td>
<td>0.5</td>
<td>18</td>
</tr>
</tbody>
</table>

* RO indicates recurrent otorrhea; HL, hearing loss; and ellipses, data not available.

The natural history of SOM in patients with PCD is unknown. In the literature, the assessment of the auditory disturbance in such patients has usually been based on small cohorts, often including children and adults. In children younger than 15 years, hearing loss appears to
be consistently associated with SOM, whereas this is not the case in adults. It is probable that in some children with PCD, SOM tends to resolve spontaneously in adulthood. The main complication that has been reported is tympanic membrane perforation. As yet, there have been no reports in patients with PCD of retraction pockets and cholesteatomas, and it is difficult to draw a conclusion as to the eventual role of PCD in the origin of the cholesteatoma in the case described herein.

Despite the limited number of cases studied, several points should be emphasized. The recurrence of SOM would appear to be very frequent (6 of 9) after type 1 tympanoplasty in children with PCD. In light of these results, can closure of a tympanic membrane perforation in children with PCD be justified? Serous otitis media is known to be associated with histopathologic changes of the tympanic membrane, and atelectasis of the graft or development of retraction pockets has been described after tympanoplasty that used temporal fascia. However, in our series, patients who underwent type 1 tympanoplasty benefited from this operation: the success rate in terms of closure of the tympanic membrane was comparable with that observed in the literature for children with no predisposing conditions, and the restrictions on bathing could probably be lifted. Preoperative otorrhea did not recur after the operation. We did not observe retraction pockets or atelectasis of the graft in the postoperative period, which included a mean follow-up of 26.8 months.

Moreover, despite the recurrence of SOM in 6 of 9 ears, speech frequency air conduction in these patients was improved, with an auditory gain of 17-dB hearing loss. This improvement was particularly appreciable in the 3 patients who had bilateral tympanic perforation. Mastoidectomy combined with type 1 tympanoplasty did not modify the anatomical and functional results in this group.

In our opinion, repair of a perforated tympanic membrane is indicated in this population when there is significant hearing loss or recurrent otorrhea. However, in the case of a small unilateral perforation with mild hearing loss and without episodes of otorrhea, we would not perform a tympanoplasty in children with PCD.

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Table 2. Preoperative and Postoperative Audiometric Testing Results in Type 1 Tympanoplasty Group

<table>
<thead>
<tr>
<th>Audimetric Measures</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>t</th>
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</thead>
<tbody>
<tr>
<td>Mean air conduction in 0.5-, 1.0-, and 2.0-kHz frequencies</td>
<td>32.77</td>
<td>16.25</td>
<td>0.002</td>
</tr>
<tr>
<td>Mean bone conduction in 0.5-, 1.0-, and 2.0-kHz frequencies</td>
<td>6.11</td>
<td>4.37</td>
<td>0.128</td>
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<td>Mean 4-kHz bone conduction</td>
<td>9.44</td>
<td>6.25</td>
<td>0.186</td>
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<tr>
<td>Mean air-bone gap in 0.5-, 1.0-, and 2.0-kHz frequencies</td>
<td>26.66</td>
<td>15.62</td>
<td>0.025</td>
</tr>
</tbody>
</table>

REFERENCES