Effect of an Intraoperative Perilymph Gusher on Cochlear Implant Performance in Children with Labyrinthine Malformations

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Background: Children with inner ear malformations often provide a special clinical challenge in that cochlear implant outcomes seem less predictable and maybe even less favorable when compared to children with normal anatomical findings.

In a recent report, we were able to demonstrate the broad range of speech perception abilities of these children when using cochlear implants. Many children with classic Mondini malformations comprising of an enlarged vestibular aqueduct (EVA) and an incompletely partitioned cochlea (IP), for example, demonstrate auditory abilities similar to children with normal labyrinthine morphology. Children with common cavity malformations (CC), on the other hand, rarely will reach meaningful auditory input beyond speech pattern perception. Likewise, cochlear implantation in ears without a radiographically visible cochlear nerve (also termed cochlear nerve deficiency, CND) will usually result in a lack of meaningful auditory skills all together.

Besides the complex audiologic features, the surgical aspect of implanting malformed inner ears remains challenging. Many cases demonstrate a more or less violent gush of perilymph fluid upon cochlear opening. While ample literature exists on how to manage these cases surgically, little is known on what effect the intraoperative encounter of a perilymph gusher might have on postoperative speech perception performance with the cochlear implant.

Aims: The aim of this report was to determine the effect of an intraoperative perilymph fluid gusher on cochlear implant speech perception performance in a large pediatric population with various types of inner ear malformations.

Table 1: Data on children with malformations grouped based on the presence or absence of an intraoperative fluid gusher

<table>
<thead>
<tr>
<th>Malformation Type</th>
<th>Presence of Gusher</th>
<th>Speech Perception Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal Labyrinthine</td>
<td>No</td>
<td>85</td>
</tr>
<tr>
<td>Normal Labyrinthine</td>
<td>Yes</td>
<td>70</td>
</tr>
</tbody>
</table>

Materials & Methods:

Patient Recruitment: 100 children with inner ear malformations; 70 had available data and were subsequently included in this study. Each child had to receive a cochlear implant in the study institution and had to have documented presence or absence of an intraoperative perilymph fluid gusher. Furthermore, adequate speech perception and production data had to be available.

Based on the intraoperative findings of an intraoperative perilymph fluid gusher, the children were assigned to two respective groups (gusher vs. non-gusher). These groups were then further subdivided based on the classification of the inner ear malformations. Both groups were compared to a control group of children who received a cochlear implant and had Connexin 26 mutations.

Surgical Management of Perilymph Fluid-Gushers: A transient facial nerve palsy approach with facial nerve monitoring was used for all surgeries. The cochlear vestibule was implanted in all instances except for CC cases where a labyrinthectomy approach was used. In some revision cases, an incomplete partitioning of the cochleovestibular nerve (IP) was used when a cochlear nerve deficiency was present.

Table 2: Imaging data for all 30 cases demonstrating an intraoperative fluid gusher

<table>
<thead>
<tr>
<th>Case Number</th>
<th>Malformation Type</th>
<th>Age (years)</th>
<th>Presence of Gusher</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal Labyrinthine</td>
<td>5</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>Normal Labyrinthine</td>
<td>7</td>
<td>No</td>
</tr>
<tr>
<td>3</td>
<td>Normal Labyrinthine</td>
<td>9</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Speech Perception Test: The Speech Perception Test (SPT) was used to evaluate the patient's speech perception performance. The test was administered when the patient had achieved at least 50% correct on the most difficult test. Performance metrics following surgery were chosen from a function of duration of implant use to show peak performance. The best score on the most difficult test are displayed visually as a function of duration of implant use to show peak performance.

Results:

Patients: 70 children (40 male, 30 female) with inner ear malformations.
- Perilymph gusher in 30 patients
- No perilymph gusher in 40 patients
- Connexin 26 positive control group: 15 patients

Present Perilymph Gusher Group (n=30):
- Associated syndromes in 11 children (36.7%)
- Joubert syndrome in 3 children (10.0%)
- Age at implantation was 73 months (45–120 months)
- Age at surgery was 1.6 years (1–5 years)
- EVA types and those children tend to have superior results across malformations, strong statistically significant correlations were found.

Speech Perception Data Across Groups: Statistical testing was performed to elucidate relevant differences. Speech perception tests were not statistically significant when comparing within malformation groups (hypoplastic + labyrinthine anomalies with or without gusher and EP-EVA cases without gusher). However, when comparing groups across malformations, strong statistically significant relationships were found.

Discussion & Conclusions: Based on the data presented herein, we can conclude that the shear presence or absence of an intraoperative perilymph fluid gusher does not seem to affect cochlear implant performance. Instead, it seems that anatomical variables such as the type of the inner ear malformation seem to affect outcomes to a much greater degree. Specifically, while children with classic Mondini malformations seem to attain excellent speech perception abilities, more severe malformations such as hypoplastic or cystic anomalies were clearly associated with poorer scores.