Current Techniques in Management of Postmeningitic Deafness in Children

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Objectives: To determine pneumococcal vaccination status of children with recent postmeningitic deafness and to review our current approach for achieving early implantation in this special population that is at significant risk for cochlear ossification.

Design: Review of imaging studies and test results.


Patients: Five children ranging in age from 15 months to 10 years who experienced recent onset of profound bilateral sensorineural hearing loss due to pneumococcal meningitis.

Interventions: All children underwent preoperative magnetic resonance imaging with 3-dimensional heavily T2-weighted steady-state free precession sequences. Four children underwent auditory steady-state response testing. All underwent bilateral cochlear implantation.

Main Outcome Measure: Degree of electrode insertion using standard surgical procedures.

Results: All children developed meningitis despite a history of pneumococcal vaccination. Complete electrode insertion in both ears was achieved.

Conclusions: Pneumococcal vaccination has reduced but not eliminated childhood deafness secondary to pneumococcal disease. Auditory steady-state response testing and 3-dimensional steady-state free precession imaging are modalities that expedite candidacy evaluation of this population. Early bilateral simultaneous implantation increases the likelihood of binaural hearing and ensures implantation of the better ear in this population of children whose course is often complicated by formation of scar tissue and ossification within the cochlea.


Since widespread use of the 7-valent pneumococcal conjugate vaccine (PCV7) began in the United States in 2001, the incidence of invasive pneumococcal disease, including meningitis, has dramatically declined.1-4 As a result, postmeningitic deafness is no longer a common cause of acquired hearing loss in childhood. However, pneumococcal meningitis continues to occur, even in healthy children who receive the recommended PCV7 vaccination series in early childhood. For this reason, it is important for cochlear implant (CI) programs to remain prepared to proceed with expeditious implantation in children recently deafened by meningitis. Timely intervention is necessary to minimize the risk of cochlear ossification, which precludes successful surgical management. At the Children’s Memorial Hospital CI program, the application of auditory steady-state response (ASSR) testing and 3-dimensional steady-state free precession (3D SSFP) magnetic resonance imaging (MRI) has been beneficial in achieving timely evaluation among CI candidates recently deafened by meningitis. In addition, it is our belief that bilateral simultaneous implantation is the treatment of choice for this special population.

Urgent evaluation in children deafened by pneumococcal meningitis is necessary because of the common occurrence of cochlear ossification. The presence of extensive ossification may limit electrode insertion and in some cases preclude even partial insertion. Special surgical approaches were developed in the past to facilitate optimal electrode placement in extensively ossified cochlea.5-8 However, because the incidence of postmeningitic deafness has declined, so has experience with these techniques. Ossification within the scala tympani typically begins near or involves the round window membrane and spreads apically. However, ossification within the scala tympani may be preceded by involvement of the semicircular canals.9 The process of ossification varies significantly in terms of
The subjects of this study were 5 children who developed postmeningitic deafness secondary to pneumococcal disease despite having received appropriate pneumococcal vaccinations. These children underwent CI between December 1, 2005, and November 30, 2007, and were the only children who underwent evaluation at our center with acquired deafness secondary to meningitis during that period. Pertinent medical history including pneumococcal vaccination status, results of auditory brainstem response (ABR)/ASSR testing and 3D SSFP MRI, degree of scala tympani obstruction, and achievement of electrode insertion constituted the main focus of review. The SSFP images were acquired on a scanner from which these sequences are known by the trademark name of FIESTA (fast imaging employing steady state acquisition; General Electric, Milwaukee, Wisconsin).

PATIENT USE OF CIs

A significant decrease in invasive pneumococcal disease has occurred since 2000, when the American Academy of Pediatrics recommended widespread use of PCV7 in the United States. However, pediatric CI centers need to remain prepared to perform expeditious evaluation in children deafened by this pathogen known to be capable of causing progressive ossification that interferes with surgical insertion of the CI electrode array. In our experience, ASSR testing and 3D SSFP MRI are useful in the candidacy evaluation of these patients and may help in facilitating successful implantation before the onset of significant cochlear ossification.

At our children’s hospital, all ABR evaluations include ASSR testing when the results reveal hearing loss in the
severe to profound range. The ASSR evaluation is advantageous in that it provides information regarding auditory thresholds in the profound range and greater frequency-specific information than ABR testing alone.13-15 A number of studies have discussed ASSR testing as an important tool in evaluating pediatric CI candidates.16-18 Our CI center has found this information to be helpful in counseling families regarding CI candidacy and in assisting the audiologist with more rapidly achieving optimal amplification, thereby shortening the hearing aid trial. Fortunately, good correlation exists between ASSR results and behavioral thresholds.13-16 In our postmeningitic patients, these advantages were especially important. Patients 1 and 2 underwent a brief hearing aid trial. However, we relied on ABR/ASSR and imaging results alone to determine CI candidacy in the other 3 cases. Although our standard evaluation protocol in infants and children includes at least a 2- to 3-month hearing aid trial in conjunction with hearing therapy, this approach was not followed for these children with recent onset of postpneumococcal deafness. A more aggressive approach was used to optimize the likelihood of full electrode insertion in both

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Time After Meningitis, d</th>
<th>Cochlear Findings</th>
<th>Time After Meningitis, d</th>
<th>Surgical Findingsa</th>
<th>Interval, db</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>38</td>
<td>Significant decrease in T2 signal bilaterally</td>
<td>41</td>
<td>Soft tissue filling inferior basal turn, extending into superior aspect of basal turn bilaterally</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>34</td>
<td>Right, minimal decreased signal in vestibules and SCCs</td>
<td>56</td>
<td>3-mm of soft-tissue obstruction bilaterally</td>
<td>22</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Left, normal signal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>6</td>
<td>Significant decrease in T2 signal bilaterally</td>
<td>12</td>
<td>Right, soft tissue filling basal turn Left, 4 mm of soft tissue</td>
<td>6</td>
</tr>
<tr>
<td>4</td>
<td>30</td>
<td>Normal T2 signal</td>
<td>43</td>
<td>Right, 3 mm of soft tissue Left, 3 mm of narrowing from ossified round window</td>
<td>13</td>
</tr>
<tr>
<td>5</td>
<td>27</td>
<td>Significant decrease in T2 signal in vestibule and SCCs bilaterally</td>
<td>32</td>
<td>4 mm of soft tissue bilaterally</td>
<td>5</td>
</tr>
</tbody>
</table>

Abbreviations: SCCs, semicircular canals; 3D SSFP, 3-dimensional steady-state free precession.

a Indicates scala tympani, with extension of the ossification beginning at the level of the round window.
b Indicates interval from magnetic resonance imaging to cochlear implantation.

Figure. Heavily T2-weighted magnetic resonance imaging with steady-state free precession sequences of the cochlea. Right (A) and left (B) cochlea demonstrate significant loss of T2 signal intensity on day 5 after onset of meningitis. Normal images (C and D) are shown for comparison.
ears in a population known to be at risk for progressive ossification.

Magnetic resonance imaging has been advocated by many as the superior modality for determining cochlear patency. Some implant surgeons use MRI exclusively for the evaluation of CI candidates because computed tomographic evaluation of the temporal bone does not, in their experience, typically provide additional information that is clinically useful. However, the sensitivity of MRI depends on the hardware and software available and the specific protocols in place to image the cochlea. At our medical center, we have found 3D SSFP scanning to be particularly helpful. Three-dimensional SSFP is a method of MRI that uses T2-weighted sequences to obtain very high contrast between fluid and solid tissue. In our experience, this imaging technique provides clinically useful information regarding cochlear architecture and patency. Steady-state free precession imaging is typically referred to by its trademarked name or acronym (FIESTA, or CISS [constructive interference steady state; Siemens, Erlangen, Germany]).

As noted in Table 2, the MRI and surgical findings regarding degree of obstruction of the basal turn were not always in agreement. It is unknown to what degree the discrepancies between the imaging and surgical findings were related to the sensitivity of the imaging vs a change in the status of the cochlea during the interval between imaging and surgical intervention. However, the children with dramatic decline in T2 signal intensity did have more significant soft-tissue obstruction of the basal turn at surgery. Children whose MRIs were consistent with patent cochlear turns had a lesser degree of obstruction at surgery.

Bilateral cochlear implantation is of special importance in children with postmeningitic deafness. Despite complete electrode insertion, 2 children (patients 1 and 3) are using only 1 device. Both of these children indicated that use of the processor in 1 of their ears is not useful despite multiple reprogramming sessions. Significant damage to the cochlear nerve by meningitis is suspected to be the underlying cause of these apparent ear differences. In these 2 cases, the preferred ear was not predictable preoperatively on the basis of imaging, audiologic testing, or intraoperative findings. Had only 1 ear undergone implantation and the second ear been addressed later, the obstruction found at the later surgery might have been more difficult to address. These 2 cases illustrate how bilateral simultaneous implantation may serve to capture the better ear in situations in which no difference between the 2 ears is evident preoperatively. Simultaneous bilateral implantation also avoids a delay that may permit development of additional ossification compromising electrode insertion.

The use of ASSR testing and 3D SSFP MRI enabled these 5 children to undergo expedited evaluation for CI candidacy. Although there was evidence of soft-tissue obstruction of the cochlea in all ears at the time of surgery, in no case was there significant cochlear ossification or mature scar tissue that might have limited insertion. Cochlea with soft-tissue obstruction in the superior aspect of the basal turn were successfully dealt with by using urologic catheters as a bougie. The HiRes 90K device was inserted, a potential advantage in the obstructed cochlea in which insertion may be challenging.

All of our patients had received pneumococcal vaccinations appropriate to their age and medical condition (Table 1). Only the child with a history of splenectomy was known to be at risk for pneumococcal disease. She developed meningitis despite daily oral penicillin prophylaxis and a 23-valent pneumococcal polysaccharide vaccine (PPV23) immunization 5 years earlier. The other 4 children had received multiple doses of PCV7 as recommended by the American Academy of Pediatrics. Three of these 4 children developed disease caused by a serotype not included in PCV7 but contained in the PPV23 vaccine (Table 1). However, these children had no risk indications to have received PPV23, and were younger than the age at which this vaccine is effective. Patient 5 experienced disease secondary to serotype 4, despite its inclusion in the PCV7 series.

A BRIEF HISTORY OF PNEUMOCOCCAL VACCINATION

When CI was in its infancy, pneumococcal meningitis was one of the most common causes of acquired postnatal deafness. At that time, only PPV23 (Pneumovax) was available to provide protection against invasive pneumococcal disease. However, this type of vaccine is not effective in children younger than 2 years, the age group most likely to experience pneumococcal meningitis. Thus, PPV23 use is limited to patients older than 2 years with known risk factors for pneumococcal disease. To address the significant morbidity and mortality brought about by pneumococcal disease in younger children, a conjugate vaccine, PCV7, was developed. It contains 7 serotypes (4, 6B, 9V, 14, 18C, 19F, and 23F) that, as a group, cause most invasive pneumococcal disease in children younger than 5 years. The PCV7 vaccination is given at 2, 4, and 6 months of age (the primary series) with an additional booster vaccination that is usually given by 24 months of age. Significant protection occurs in most infants after even the first vaccination of the primary series. The American Academy of Pediatrics and Centers for Disease Control and Prevention recommended widespread use of PCV7 in mid-2000. Implementation of their guidelines resulted in a significant decline in invasive pneumococcal disease, including meningitis.

GENERAL IMPLICATIONS FOR MENINGITIS PREVENTION BY PEDIATRIC CI CENTERS

Our experience with acquired deafness secondary to pneumococcal disease in vaccinated children has heightened our CI team’s awareness of the importance and potential limitations of pneumococcal vaccinations in our general pediatric CI population. The possible association between bacterial meningitis and the presence of a cochlear implant was recognized in June 2002. Most of the cases identified at that time had occurred in the pediatric population and were secondary to pneumococcal disease. A major risk factor was determined to be the presence of a positioner (Advanced Bionics Corporation) that was placed adjacent to the electrode array. The positioner was
removed from the market in 2002. Other significant risk factors are the presence of cerebrospinal fluid leak and cochlear malformations. In 2003, the Centers for Disease Control and Prevention released vaccination guidelines to help prevent meningitis in CI recipients. These guidelines recommend that all pediatric candidates and recipients younger than 5 years receive PCV7 according to the high-risk schedule recommended by the American Academy of Pediatrics. It was also recommended that children 2 years or older receive PPV23. Study of CI recipients revealed additional risk factors for meningitis beyond the presence of the implant itself, including cerebrospinal fluid leak at the time of implantation and the presence of cochlear malformations, a ventriculoperitoneal shunt, a positioner, and otorrhea. Although a follow-up study of the incidence of meningitis in CI recipients showed a decline beyond 24 months after implantation, there is reason for continued concern. Several cases of pneumococcal meningitis in children with CIs who were eligible for but had not yet received PPV23 have been reported.

At our CI center, we ensure that CI candidates receive vaccinations against hemophilus influenza B and pneumococcus as recommended by the Centers for Disease Control and Prevention. Before implantation, we prefer that children younger than 2 years receive the PCV7 primary series to the degree that would be appropriate on the basis of age. Because most new CI candidates we evaluate have already received PCV7 at their primary care physician’s office as part of their routine childhood vaccination schedule, the need to vaccinate young children rarely affects the timing of surgery. In the event of an unvaccinated child, the issue of delaying surgery until all age-appropriate PCV7 vaccinations have been completed needs to be determined on a case-by-case basis. The child’s overall medical condition and risk of meningitis must be weighed against the benefits of younger age at implantation. We also place special emphasis on children who underwent CI before age 2 years receiving their PPV23 as soon as possible after their second birthday. This latter recommendation is important because PPV23 has the potential to provide children with protection against an additional 16 pneumococcal serotypes beyond the 7 that are part of the conjugate vaccine.

**CONCLUSIONS**

Despite the dramatic decline in invasive pneumococcal disease subsequent to widespread use of PCV7 in the United States, pneumococcal meningitis as a cause of deafness has not been eliminated. Indeed, with more than 90 pneumococcal serotypes in existence, eradication is not expected even if current vaccine protocols continue to successfully limit the incidence of invasive disease. Therefore, pediatric CI programs need to remain prepared to evaluate and perform implantation in this special population. In our experience, more information is obtained and clinical management facilitated by the addition of ASSR to the ABR evaluation. Likewise, use of 3D SSFP MRI provides useful information about significant changes in cochlear patency before the onset of new bone formation within the cochlea. In light of the unpredictable nature of post- meningitic ossification, we recommend that CI surgeons consider bilateral simultaneous implantations to increase the likelihood of successful electrode array insertions, thereby preserving the potential for these children to achieve useful binaural hearing.

**Submitted for Publication:** May 14, 2009; final revision received March 22, 2010; accepted April 19, 2010.

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**Author Contributions:** Dr Young had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

**Study concept and design:** Young. Acquisition of data: Young. Analysis and interpretation of data: Young and Tan. Drafting of the manuscript: Young and Tan. Critical revision of the manuscript for important intellectual content: Young and Tan. Administrative, technical, and material support: Young.

**Financial Disclosure:** Dr Young serves on the medical advisory boards of Cochlear Americas and Advanced Bionics Corporation. Dr Tan serves on the medical advisory board and speaker’s bureau of Wyeth Vaccines.

**Funding/Support:** This study was supported in part by the Lillian S. Wells Foundation.

**Role of the Sponsor:** The sponsoring institution was not involved in the study design, data, and/or manuscript aspects of this study.

**REFERENCES**


