Osteogenesis Imperfecta and Hearing Loss in the Pediatric Population

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ABSTRACT

Osteogenesis Imperfecta (OI) has long been associated with hearing loss. This hearing loss is thought to occur in the second to third decade of life in the majority of patients. Current data are not conclusive on when to start screening children with OI for hearing loss and how often to repeat screening. This presented a significant question for our investigators. Our goal was to determine the prevalence and incidence of hearing loss in the pediatric OI population and at what age hearing screening begin in these children.

RESULTS

- Data was collected from 143 patients
- 127 patients had PTA or SFA available
- Average age of initial screening was 5.1 years (SD=4.8)
- Prevalence was 24% (n=30)
- Incidence was 9% (n=10)
- 21 patients had PTA data available (see Table 1) to determine type of hearing loss
- Average age of onset for hearing loss 5.8 years (SD=4.9)
- Most children had a mild hearing loss (see Table 2)

DISCUSSION

In our study, we found that the majority of patients had a conductive hearing loss. This is in agreement with other studies that have also found conductive hearing loss as the dominant type, despite age.1-5,7 The prevalence of hearing loss in our study (24%), which falls in the middle of the range of previous studies (see Table 3), was 9% (n=10). In their study, they looked at 45 patients and found hearing loss in three children, ages seven, eleven, and fifteen. The average age of onset for hearing loss in our population was 5.8 years.

In our search for recommendations on when to start screening children with OI for hearing loss, only one article was found that gave clear guidelines. Kuurila et al. recommended that screening should start at ten years of age and then be repeated every three years. In our study, the prevalence of hearing loss was 24% and the incidence of 9%. The mean age of onset for hearing loss was 5.8 years. Compared to the previous study, we would recommend screening children with OI at the time of diagnosis with the disease. These children should also receive routine scheduled follow up.

CONCLUSIONS

- In our study the prevalence of hearing loss was 24% and the incidence of 9%
- The majority of patients in our study had a conductive hearing loss
- The average age of onset was 5.8 years
- We support hearing screening in children at the time of diagnosis with OI and routine scheduled follow up

METHODS AND MATERIALS

- Retrospective chart review
- Study participants: Patients 19 years or younger that attended one or more OI clinic at Children’s Hospital
- Data collected:
  - Pure tone audiometry (PTA)
  - Sound field audiometry (SFA)
  - Tympanograms
  - Otoacoustic emissions (OAEs)
  - Type of hearing loss
  - Severity of hearing loss
  - Only patients with information in the form of PTA or SFA were included in the statistical analysis

REFERENCES


CONTACT

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Table 1. Type of Hearing Loss.

<table>
<thead>
<tr>
<th>Type</th>
<th>PTA</th>
<th>SFA</th>
<th>Combined</th>
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</thead>
<tbody>
<tr>
<td>Right (n=18)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CD</td>
<td>12%</td>
<td>14%</td>
<td>18%</td>
</tr>
<tr>
<td>SN</td>
<td>7 (44%)</td>
<td>7 (39%)</td>
<td></td>
</tr>
<tr>
<td>Mixed</td>
<td>2 (11%)</td>
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</tbody>
</table>

Table 2. Severity of Hearing Loss.

<table>
<thead>
<tr>
<th>Type</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
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</thead>
<tbody>
<tr>
<td>Right (n=18)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Initial Evaluation</td>
<td>8/11</td>
<td>12/18</td>
<td>10/18</td>
</tr>
<tr>
<td>OAE</td>
<td>8/11</td>
<td>12/18</td>
<td>10/18</td>
</tr>
<tr>
<td>Combined</td>
<td>8/11</td>
<td>12/18</td>
<td>10/18</td>
</tr>
</tbody>
</table>

Table 3. Comparison with Previous Studies.