**Clinical Focus**

**Effect of Amplification on Speech and Language in Children With Aural Atresia**

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**Purpose:** There currently is no guideline regarding amplification or verification for children with aural atresia. This population, with the absence of the ear canal, has obvious congenital hearing loss. Research suggests that delayed treatment for hearing loss can result in speech and language impairments, including poor performance in school. In this study we examined the relationship between amplification and emerging speech and language development in children with aural atresia.

**Method:** Subjects included children, 3 to 6 years of age, with conductive hearing loss due to atresia. Clinical evaluations were used to determine relationships between amplification and speech and language development. Subjects underwent an audiology exam, including pure-tone or warble-tone air and bone conduction using play audiometry techniques, and a speech and language evaluation assessing language and vocabulary skills.

**Results:** Subjects who were fitted before 1 year of age showed better compliance with aid use and exhibited fewer delays in speech and language development. Children with right-sided atresia had significantly greater speech and language delays when compared with age norms from standardized tests.

**Conclusion:** Our findings highlight a relationship between side of atresia, time of first amplification, compliance in aid use, and speech and language abilities.

Hearing loss, whether it is conductive or sensorineural, has the potential to significantly affect a child’s development and overall quality of life. Although it is well documented that children with hearing loss have more delays in speech development and education, it is not standard practice in many cases to provide amplification or other intervention until delays are evident. The guidelines put forth by the American Academy of Audiology (Ching et al., 2013) state that hearing aid amplification or other intervention in children with unilateral hearing loss, mild sensorineural loss, or conductive hearing loss should be made on a case-by-case basis, taking into consideration a variety of factors such as child and family preference, communication abilities, and educational success (McKay, 2010). However, delaying amplification until there is an indication of communication delays or educational problems may diminish the effectiveness of potential interventions and force the child into playing catch-up. Delays in speech and language abilities have been identified in several studies of children with hearing loss ranging from 10 months to 20 years of age (Borg et al., 2002; Klee & Davis-Dansky, 1986; Lieu, Tye-Murray, Karzon, & Piccirillo, 2010; Peckham & Sheridan, 1976). Overall academic performance has been found to be lower in children with hearing loss compared with children with normal hearing (Hayiou-Thomas, Harlaar, Dale, & Plomin, 2010; Lieu, 2004). In addition, one study found that the academic performance of children with unilateral hearing loss was worse than that of children with bilateral hearing loss, possibly due to the inconsistent intervention and amplification in children with unilateral hearing loss (Most, 2004) resulting from a lack of clinician guidelines. It has been suggested that children with hearing loss who begin both audiological and speech-language therapy services at an early age may develop language on par with that of their peers without hearing loss (American Speech-Language-Hearing Association, 2008).

This preliminary investigation focused on amplification for children with unilateral and bilateral aural atresia or microtia. Microtia (Latin for “little ear”) is a congenital deformity affecting the outer ear (pinna) in which the ear does not fully develop during the first trimester of pregnancy. A microtia ear is often smaller in size, can have a peanut-shaped appearance or only a small nub or lobe, or can be completely absent at birth. Microtia can affect one ear (unilateral) or both ears (bilateral). Microtia occurs in one out of every 6,000 to 12,000 births. The right ear is more commonly affected. Microtia is often accompanied by atresia.
(also known as aural atresia), which is the absence or closure of the external auditory ear canal. The middle ear bones (incus, stapes, and malleus) may be malformed, including the narrowing of the ear canal (known as canal stenosis). Atresia is Latin for “absence of an opening.” The prevalence of unilateral atresia or microtia is significantly increased in Hispanic communities (Ramadhani et al., 2009). This diagnosis is prevalent in Southern California’s large Hispanic population. An epidemiologic study in California with data from 1989 to 1997 found the prevalence of atresia to be 2.5 for every 10,000 live births (Shaw, Carmichael, Kaidarova, & Harris, 2004). Compared with Caucasians, the risk of aural atresia is three times higher in Asians, and seven times higher in Hispanics. In addition to racial and ethnic differences, lower maternal education was associated with an increased risk of aural atresia in this data set (Shaw et al., 2004).

Recent studies have begun to examine children with aural atresia more closely. However, there remain no specific clinical guidelines (i.e., when intervention should be started, whether amplification or assistive devices should be used, and what type of device is most effective) concerning intervention for unilateral conductive and sensorineural hearing loss. Intervention may consist of conventional hearing aids, Frequency Modulation (FM) systems, Contralateral Routing of Signal (CROS) hearing aids, transcranial amplification, and bone-conduction devices, which are available in both surgical and nonsurgical options (McKay, 2010; McKay, Gravel, & Tharpe, 2008). In a population of school-age children with aural atresia, only 12.5% used a hearing aid and 32% used an FM system, whereas 65% needed some resource such as speech therapy or were on an individualized education plan (Kesser, Krook, & Gray, 2013). A recent study found high rates of speech therapy in children with aural atresia (Jensen, Grames, & Lieu, 2013): Eighty-six percent of those with bilateral atresia and 43% of those with unilateral atresia had speech therapy. It is interesting to note that a higher percentage of children with right-side aural atresia had greater problems in school compared with children with left-side or bilateral atresia (Jensen et al., 2013). Although some data on aural atresia and performance in school-age children are emerging, it remains unclear when delays are evident and whether children younger than 6 years of age already show speech and language delays. The goal of this study was to investigate how interventions influence outcomes for children with aural atresia. Using current patients in an audiology clinic, we aimed to gain preliminary insight into whether the type of intervention, time of first intervention, and compliance with wearing hearing aids and assistive devices can be correlated with speech and language abilities of children with conductive hearing loss due to atresia.

Method

Subjects

Children between 3 and 6 years of age with unilateral or bilateral permanent conductive hearing loss due to aural atresia were included in this study. This population was a convenience sample identified through a screening of current and previous patients treated at Casa Colina Hospital and Centers for Healthcare in Pomona, California. There was no requirement for amplification use; however, children were excluded if they had any additional comorbidities, acquired or developmental, such as Down syndrome, cerebral palsy, or autism. After potential subjects were identified ($n = 40$), the parents of the potential subjects were contacted and informed of the study. Informed consent was completed prior to enrollment, and this study was done in accordance with required ethical standards.

Study Design

Upon enrollment in the study, each child completed a baseline audiology exam and speech evaluation. A parent or legal guardian for each subject was asked to complete the Children’s Outcomes Worksheet (COW; Williams, 2004), which is designed to assess a child’s needs and abilities prior to and after amplification. The COW provides a simple individualized assessment of a child’s needs and a subsequent assessment of how well the rehabilitation process addresses those needs. The COW determines whether the fitting process has resulted in change and assesses the child’s ability after use of the new hearing instrument.

The degree of change and the child’s ability in a number of different situations (e.g., in a car, during a conversation, in a large group) are assessed by both the parent and the child.

Audiology Evaluation

Subjects completed an audiology exam, which included pure-tone or warble-tone air and bone conduction using play audiometry techniques to ensure there was no underlying sensorineural component to the hearing loss. All speech testing was completed using a live voice in the child’s primary language. Speech thresholds for each ear were recorded using picture identification of spondaic words. When vocabulary was good enough, word recognition was tested using Word Intelligibility by Picture Identification (Ross & Lerman, 1979). Middle ear function was measured using tympanometry in the non-atretic ear to ensure that no other confounding conductive hearing component was present. In subjects currently using an amplification device, an evaluation of the device was completed. This evaluation included sound field warble-tone thresholds and sound field speech thresholds with appropriate masking of the unaided ear. For digital instruments, devices were connected to the manufacturer software for setting validation and data logging. No objective verification equipment for bone-conduction devices is currently available.

Speech Evaluation

All subjects underwent a speech and language evaluation in their primary language (English or Spanish) using the Preschool Language Scale–Fourth Edition (PLS-4; Zimmerman, Steiner, & Pond, 2002) to assess language
skills and the Expressive and Receptive One-Word Picture Vocabulary Tests (EOWPVT and ROWPVT, respectively; Brownell, 2000) to assess vocabulary skills. The PLS-4 (English or Spanish) screens for a broad spectrum of language skills in young children in less than 10 min. The EOWPVT and ROWPVT (Spanish bilingual and English) are individually administered, norm-referenced assessments of how well persons ages 2 years 0 months to over 80 years can name the objects, actions, or concepts presented in full-color pictures. The test consists of items presented in a developmental sequence (based on the 2010 normative sample) that reflects the concepts with which children currently have experience through home, school, or media. All speech and language testing was performed by a licensed speech pathologist in the child’s primary language. Both a standardized T score and an age-equivalency score in months were determined from these assessments, providing a comparison between our population and the age norms from standardized tests. Delays in speech and language were determined by taking the mean of the three assessments’ age-equivalency scores. We did not note any differences between the scores on the three assessments; each child was either consistently delayed or typically developing. If a child was delayed more than 6 months from his or her current age, he or she was classified as having delays in speech and language. Children with scores ranging from 6 to 12 months behind their age were classified as having delays in speech and language. Children with scores ranging from 6 to 12 months behind their age were classified as having moderate delays, and those with delays greater than 12 months from their current age were classified as having severe delays.

Results

The study population consisted of 16 subjects (mean age = 4.2 years; 64% boys, 36% girls; 87% Hispanic) with conductive hearing loss due to unilateral (n = 10) or bilateral (n = 6) atresia. The average time between birth and first amplification was just under 2 years; however, it ranged from 3.5 months to 6 years (see Table 1). At the time of the evaluation for the study, all subjects had a bone-conduction average threshold of 3.14 dB (see Table 2). The pure-tone bone-conduction average threshold was 3.14 dB on the side with atresia and 8.3 dB for air on the non-atretic side, which represents normal hearing. In all children with unilateral atresia, hearing was normal on the non-atretic side for subjects with unilateral atresia. Parents reported the average use of the amplification device to be 5 hours/day, which matched the average use recorded with data logging.

When comparing individuals with unilateral and bilateral aural atresia, we observed that subjects with bilateral atresia were fitted with devices at a younger age compared with subjects with unilateral atresia (see Figure 1A; t = 2.3, df = 13). There was no significant difference in age between the subjects with bilateral and unilateral atresia. Data also showed that the daily use of devices was significantly longer in subjects with bilateral atresia than in subjects with unilateral atresia (see Figure 1B; p = .003, t = −3.7, df = 13). Parents perceived greater improvement in response to sounds as measured by the COW in subjects with bilateral atresia versus subjects with unilateral atresia; however, parents of subjects with bilateral atresia did not perceive a greater difference in their child’s ability to act on the sounds in the environment compared with subjects with unilateral atresia once aided (see Figure 1C; p = .047, t = −2.26, df = 13). Further, no significant difference in speech and language abilities (as measured with the PLS-4, EOWPVT, and ROWPVT) was observed between subjects with bilateral and unilateral atresia. However, the subjects with bilateral atresia did show a trend toward exhibiting greater speech and language delays (see Figure 1D). Although the majority of the population was Spanish speaking, we did observe a nonsignificant trend toward greater delays in the Spanish-speaking children.

On the basis of age-adjusted test scores, speech and language delays (mean of the three tests) were analyzed by grouping children into three groups: (a) typical development, (b) mild delay (delayed by 6–12 months), and (c) severe developmental delay (delayed by more than 12 months). We found no significant difference in delays by age, although the children with severe delays tended to be the oldest (see Figure 2A), an effect that is not unexpected due to the normative range becoming smaller with increasing age. This effect was consistent for both children with unilateral atresia and children with bilateral atresia. Children with mild and severe delays wore their aids on average 2 hr/day more compared with children with typical development (see Figure 2B). We also observed that children who were fitted with their first amplification device later exhibited more severe speech and language delays and were less compliant in using their aid, although again these effects were not significant (see Figure 2C). It is interesting to note that in

Table 1. Subject characteristics.

<table>
<thead>
<tr>
<th>Group</th>
<th>Age at enrollment (yrs)</th>
<th>Male subjects</th>
<th>Female subjects</th>
<th>L side of loss</th>
<th>R side of loss</th>
<th>Time from birth to first fitting (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M ± SD</td>
<td>Range</td>
<td>#</td>
<td>%</td>
<td>#</td>
<td>%</td>
</tr>
<tr>
<td>All (n = 16)</td>
<td>4.2 ± 0.9</td>
<td>3–6</td>
<td>10</td>
<td>63</td>
<td>6</td>
<td>37</td>
</tr>
<tr>
<td>Unilateral atresia (n = 10)</td>
<td>4.4 ± 1.0</td>
<td>3–6</td>
<td>5</td>
<td>50</td>
<td>5</td>
<td>50</td>
</tr>
<tr>
<td>Bilateral atresia (n = 6)</td>
<td>3.9 ± 0.9</td>
<td>3–5</td>
<td>5</td>
<td>83</td>
<td>1</td>
<td>17</td>
</tr>
</tbody>
</table>

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children with developmental delays, those with right-sided atresia displayed greater delays (see Figure 2D; $p = .035$, $t = -2.42$, $df = 5$). Among the children with right-sided atresia, the children with typical development were fitted on average 1 year earlier than the children with delays. Although our sample size is small, these data suggest that fitting children earlier may prevent some delays, especially in children with right-sided atresia, who appear to be more vulnerable than those with left-sided atresia.

**Discussion**

Atresia is a hearing loss that is evident at birth. However, even with early identification and intervention practices, many children with atresia go without help for many years. We sought to begin to empirically address this issue by studying early intervention and the benefits of amplification on speech and language development. This study investigated the effect of hearing loss due to unilateral

Table 2. Subject amplification and hearing loss characteristics.

<table>
<thead>
<tr>
<th>Variable</th>
<th>No.</th>
<th>%</th>
<th>dB</th>
<th>Range</th>
<th>$M \pm SD$</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type of amplification device</strong></td>
<td></td>
<td></td>
<td></td>
<td>All</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Baha Divino</td>
<td>4</td>
<td>25</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Baha 3 BP100</td>
<td>5</td>
<td>31</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oticon Pronto Pro</td>
<td>4</td>
<td>25</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other bone conduction devices</td>
<td>3</td>
<td>19</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>PTA</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bone: Atretic side (masked)</td>
<td>3.14</td>
<td></td>
<td>-4.00 to 20.00</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Air: Nonatretic side</td>
<td>8.30</td>
<td></td>
<td>2.00 to 15.00</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aided</td>
<td>22.30</td>
<td></td>
<td>10.00 to 37.00</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Average daily use in hours</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Per data logging ($n = 5$)</td>
<td>5.3 ± 2.9</td>
<td></td>
<td>2.5 ± 1.3 ($n = 4$)</td>
<td>8 ($n = 1$)</td>
<td></td>
</tr>
<tr>
<td>Per parent report ($n = 10$)</td>
<td>5.1 ± 2.9</td>
<td></td>
<td>7.5 ± 2.7 ($n = 5$)</td>
<td>4.5 ± 1.7 ($n = 5$)</td>
<td></td>
</tr>
</tbody>
</table>

Note. Daily aid use was not recorded by one participant. PTA = pure-tone average.

**Figure 1.** Differences between subjects with unilateral atresia and bilateral atresia. Error bars represent standard error of the mean. COW = Children’s Outcomes Worksheet; PLS-4 = Preschool Language Scale–Fourth Edition; ROWPVT = Receptive One-Word Picture Vocabulary Test; EOWPVT = Expressive One-Word Picture Vocabulary Test.
and bilateral atresia on speech and language development. This population of children with atresia has become of particular interest to our clinic because of the large number of children with atresia or microtia referred to our audiology clinic by the newborn screening program. The lack of professional guidelines for testing and intervention has led to concern about making sure we are helping these children achieve the best developmental outcomes. Our goal was to look for relationships between amplification and speech and language development. Even the most recent pediatric amplification guidelines released by the American Academy of Audiology have no clinical guideline for amplification in children with aural atresia (Ching et al., 2013). Therefore, most interventions occur when the first delays in speech and language are observed. Our study suggests that, once aided, children with bilateral atresia display the same delays as children with unilateral atresia. We did not address the benefit of bilateral versus unilateral fitting; however, current research has shown a significant benefit in sound localization with bilateral amplification. In our clinic, we have made it policy to fit children with bilateral and unilateral atresia as young as possible. It is not in a child’s best interest to wait for delays to occur before treatment when intervention options may help prevent or minimize future educational and developmental problems.

The findings of the present study indicate a relationship between developmental delays and the time before first amplification. However, the small sample size and other uncontrolled variables limit the interpretation. The normative range for speech and language development is much broader in younger children than in older children, which may explain why delays are more apparent as children get older. We also found an interesting sidedness effect in children with right-sided atresia whereby children who developed normal speech and language were fitted 1 year earlier than children with delays. The right ear is typically the dominant ear for processing speech information. The early presentation of auditory information through bone-conduction amplification to the atretic right side may explain why these children were developing more typically compared with the children with delays. The normal development of these auditory pathways is essential to learning and success in the classroom. It has long been established that right-ear information to the auditory system is necessary for normal processing of auditory information in children. We also found that the earlier the children were fitted, the more compliant they were in wearing the devices. It stands to reason that children fitted at a young age become accustomed to the use of the band and device, which must be worn fairly tight and can be uncomfortable. Older children show greater resistance to the devices and ultimately demonstrate less use and less benefit.

Although preliminary, this type of investigation is a starting point in collecting information to establish best practices regarding early intervention in children with atresia. Although children in this population spend considerable time...
in surgery for reconstruction and repair of the facial and aural structures, the importance of hearing is often forgotten until the children are noticeably delayed in school and social development. A larger study is warranted to more clearly define the relationships between amplification and speech and language delays. Recommendations regarding interventions and rehabilitation of this population can then follow. In addition, although the pediatric amplification guidelines from the American Academy of Audiology list the use of bone-conduction devices for atresia (Ching et al., 2013), there is no protocol for objective verification of the availability of speech information for the child using the device (e.g., speech mapping). Further research in this area is required to provide assurance that we are providing optimal speech information to these children’s developing ears and brains.

Acknowledgments

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References


