Effect of amplification on speech and language in children with aural atresia

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Casa Colina Hospital and Centers for Healthcare

- Free standing Acute Rehabilitation Hospital in Pomona California.

- Outpatient services with specialist physicians, specialized therapists (vestibular, pool, wheelchair seating)

- Audiology Program (started in 2002) is a part of the Outpatient services and works closely with inpatient, outpatient, physical therapy, occupational therapy, speech therapy, and children services.
Why the interest in children with Microtia/Atresia

- Newborn Hearing Screening Program
- Large population of children with microtia/atresia in the inland empire.
- Lack of clinical guidelines for testing, and amplifying these children
- The push from early start to provide help for these children.
Presentation Outline

• Background: Why this subject should be studied

• Hypothesis / Objectives: What we were hoping to discover.

• Study Design: Who did we study?

• Results: What the results suggests.

• Conclusions / Clinical Implications: What should we take from this study.

• What is next? My challenge to the Audiology community.
Aural Atresia / Microtia

• Microtia
  – congenital deformity affecting the outer ear (pinna)
  – ear does not fully develop during the first trimester of pregnancy
  – ear may be smaller in size, have a peanut shape, a small nub or lobe, or be completely absent at birth
  – can affect one ear (unilateral) or both ears (bilateral)
  – occurs in every 1 out of 6,000 to 12,000 births
  – the right ear is more commonly affected

• Aural atresia
  – often associated with microtia
  – the absence or closure of the external auditory ear canal
  – malformation of the middle ear bones (incus, stapes, and malleus) including the narrowing of the ear canal (stenosis)
  – may arise from problems in the fetus development rather than genetic factors
Aural Atresia Epidemiology

• The prevalence of unilateral atresia/microtia is significantly higher in Hispanic communities (Ramadhani et al., 2009).

• The large Hispanic population in southern California makes atresia a prevalent diagnosis (Shaw, Carmichael, Kaidarova, & Harris, 2004)
  – 0.67 versus 2.5 for every 10,000 live births in Hispanic population

• Lower maternal education is also associated with an increased risk of aural atresia in some studies (Shaw et al., 2004)
Hearing Loss and Language Development

- Hearing loss has the potential to significantly affect a child’s receptive and expressive vocabulary.

- Such delays in speech and language abilities cause learning deficits.

- Children with hearing loss have more difficulties with social skills.

- Children with right unilateral hearing loss are more at risk for difficulties with language tasks and verbal cognitive skills.
Unilateral Hearing Loss and Academic Performance

• Overall academic performance has been found to be lower in children with UHL when compared to children with normal hearing

• Children with UHL have been shown to have worse academic performance than children with bilateral hearing loss

• Children with right unilateral hearing loss repeat grades more often and have poorer performance on verbal tests than those with left ear hearing loss
Atresia and Language Development

- High rates of speech therapy are identified in children with aural atresia
  - 86% of those with bilateral atresia had speech therapy
  - 43% of those with unilateral atresia had speech therapy

- A higher percent of children with right-sided atresia reported greater problems in school than those with left-sided atresia or bilateral atresia

- In school age children with aural atresia
  - 12.5% were using a hearing aid
  - 32% were using a FM system
  - 65% were reported as needing some resource i.e., speech therapy, or were on an Individualized Education Plan
Clinical Guidelines for Amplification

- Despite the research 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 2013 guidelines put forth by the American Academy of Audiology state that hearing aid amplification or other intervention in children with unilateral hearing loss (UHL), sensorineural loss (SNL) or conductive hearing loss, should be made on a case to case basis taking into consideration a variety of factors including child and family preference as well as communication abilities and educational success.

- The guidelines also state that we need to use reliable and valid measures of a child's progress to assess early intervention goals in speech and language development.
Does it make sense to wait

• Waiting on amplification until there is an indication of communication delays or educational problems may diminish the effectiveness of potential interventions.

• It has been suggested that children with hearing loss, who begin services early, both audiological and speech / language therapy may develop language on par with their peers without hearing loss.

• Currently, there remains no specific clinical guidelines concerning intervention for conductive and sensorineural unilateral hearing loss,
  – when the intervention should be started
  – if amplification or assistive device should be used
  – what type of device is most effective
Amplification

- There are several types of hearing assistive devices in addition to conventional hearing aids that may be considered including,
  - FM systems
  - CROS hearing aids
  - Transcranial amplification (both surgical and non-surgical options)
    - BAHA/Ponto
    - Sofono
Hypothesis

• The goal of this study was to investigate the effect of amplification through bone conduction on speech and language development for children with aural atresia.

• Using current atresia patients in a single Audiology clinic we studied speech and language development as it related to:
  – the time of first intervention
  – the compliance with wearing hearing aids and assistive devices
  – Differences in right verses left sided atresia
  – Differences in Bilateral verses unilateral atresia
Study Participants

Inclusion criteria
• Male and female
• 3 and 6 years of age
• unilateral or bilateral conductive hearing loss due to aural atresia

Exclusion criteria
Participants were excluded from the study if they had any additional co-morbidities either acquired or developmental in origin, such as Down Syndrome, cerebral palsy, or autism.
Study Design: Audiology Evaluation

- Participants completed an audiology exam which included,
  - an analysis of pure tone air and bone conduction
  - speech thresholds for each ear when possible using picture identification
  - word recognition using Word Identification by Picture Identification
- In participants currently using an amplification device
  - an evaluation of the device was completed which included,
    - sound field warble tone threshold
    - speech thresholds with masking to the unaided ear in unilateral cases
    - computer analysis of device when possible (output and data logging)
Study Design: Speech Evaluation

- All participants underwent a speech and language evaluation conducted by a speech pathologist in the child's primary language (English or Spanish) using,
  - the Preschool Language Scale 4 to assess language skills
  - the Expressive and Receptive One-Word Picture Vocabulary Tests (EOWPVT, ROWPVT) for vocabulary skills
Study Design: Children’s Outcome Worksheet

• A parent or legal guardian for each subject was asked to complete the Children’s Outcomes Worksheet (COW).

• The COW was 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 subsequent assessment of how well the rehabilitation process addresses those needs.

• The COW determines if the fitting process resulted in a change in hearing ability and what is the child’s ability level when aided.
## Results: Participant Characteristics

<table>
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<tr>
<th>Participant Characteristics</th>
<th>All Subjects (n=16)</th>
<th>Unilateral (n=10)</th>
<th>Bilateral (n=6)</th>
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<tbody>
<tr>
<td>Age at enrollment</td>
<td></td>
<td></td>
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<tr>
<td>Mean ± SD (years)</td>
<td>4.2 ± 0.9 years</td>
<td>4.4 ± 1.0 years</td>
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<tr>
<td>Range</td>
<td>3 – 6 years</td>
<td>3 – 6 years</td>
<td>3 – 5 years</td>
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<tr>
<td>Male</td>
<td>10 (63%)</td>
<td>5 (50%)</td>
<td>5 (83%)</td>
</tr>
<tr>
<td>Female</td>
<td>6 (37%)</td>
<td>5 (50%)</td>
<td>1 (17%)</td>
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<td>Side of loss</td>
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<td></td>
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<tr>
<td>Left side</td>
<td>5 (50%)</td>
<td>5 (50%)</td>
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<tr>
<td>Right side</td>
<td>5 (50%)</td>
<td>5 (50%)</td>
<td>N/A</td>
</tr>
<tr>
<td>Time from birth to 1st fitting</td>
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<td></td>
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<tr>
<td>Mean ± SD (months)</td>
<td>21.4 ± 19 mo</td>
<td>26.5 ± 20 mo</td>
<td>12.8 ± 17 mo</td>
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<tr>
<td>Range</td>
<td>3.5 – 73.4 mo</td>
<td>5.6 – 73.4 mo</td>
<td>3.5 – 47.7 mo</td>
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the COW
the Child’s Needs

Name: ________________________________

Audiologist: ____________________________

Date: 1. Needs established ____________________ 2. Outcome assessed ________

SPECIFIC NEEDS

Indicate Order of Significance

Conversation with group in noise
Specific Situation:

Hearing children in the classroom
Specific Situation:

Hearing family in home
Specific Situation:

Degree of Change
"Because of the new hearing instrument, I now hear..."

Ability with hearing instrument
"I can hear satisfactory..."

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<th>Very Poor</th>
<th>No Difference</th>
<th>Slightly Better</th>
<th>Better</th>
<th>Much Better</th>
<th>SCORE</th>
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When comparing individuals with unilateral or bilateral aural atresia we observed that bilateral subjects on average were first fit with devices at a younger age than unilateral subjects.
Bilateral subjects report/log significantly longer daily use than unilateral subjects.
Parents perceived greater improvement in response to sounds as measured by the COW in bilateral subjects versus unilateral subjects; however, parents did not perceive a difference in their child’s ability to act on the sounds in the environment.
• There were no significant difference in speech and language abilities (as measured with the PLS-4, EOWPVT and ROWPVT) between bilateral and unilateral subjects.

• The bilateral participants did show a trend towards exhibiting greater speech and language delays.
When speech and language delays (mean of the 3 tests) were analyzed according to severity we found that those with severe delays were the oldest,
Children with mild and severe delays wear their aids on average more than the typically developing subjects.
Speech and Language Delays

Children who are fit with their first amplification device later exhibit more severe speech and language delays.
• Interestingly, among children with developmental delays those with right-sided atresia displayed greater delays.

• When looking at just the children with right sided atresia, typically developing children were fit on average one year earlier.
Conclusions

- Our study suggests that once aided, children with bilateral atresia display the same delays as those with unilateral atresia.

- The earlier the children were fit, the more compliant they were at wearing the devices than children fit later.

- It goes to reason that children fit young will become accustomed to the use of the band since the 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.
Conclusions

• While the sample size is small this data suggests that fitting children earlier may prevent some delays especially with right-sided atresia
  • Right-sided children who developed normal speech and language were fit one year earlier than the right-sided children with delays
  • It is the right ear that is typically the dominant ear for processing speech information.
  • It goes to reason that hearing loss on the right could lead to more significant speech and language delays.
  • The normal development of these auditory pathways is essential to learning and success in the classroom.
Clinical Implications

• Findings from the present study are the beginnings of an attempt to help collect information to establish best practices regarding early intervention of children with atresia.

• This population will spend considerable time in surgery for reconstruction and repair of their facial and aural structures while the importance of hearing is often forgotten until they are noticeably delayed in school and social development.

• Since there is no research to verify the benefit of early fitting and the high cost of the device, many clinics choose not fit until the child is older.

• If we are fitting children with ears at very young age why not the atresia children.
Our Clinic Policy

- We have made it a policy to fit children with bilateral and unilateral atresia as young as possible.

- Children with unilateral hearing loss have problems in school, with behavior, speech development and academic success.

- In our opinion, it is not a child’s best interest to wait for delays to occur before treating when there are intervention options which may help to prevent or minimize future educational and developmental problems.

- Findings from the present study are the beginnings of an attempt to help collect information to establish best practices regarding early intervention of children with atresia.
Need for further research

• There is no objective verification of the availability of speech information for the child using the device (such as speech mapping).

• Further research in this area should include
  – attention to provide assurances that we are providing optimal speech information to these children’s developing ears and brain.
  – verification of the ability to localize sounds presented by bone conduction both unilaterally and bilaterally.
Binaural hearing ability with bone conduction stimulation in normal hearing subjects
M. Seltooni, E. Maki-Torkko, S. Stenfelt

• Conclusion:
  – Spatial release from masking in a stationary noise with speech targets was around 5 dB with BC mastoid application
  – Patients with bilateral conductive impairments and good cochlea's should be fitted bilaterally.
  – We need BCHAs that are adapted for binaural application
THANK YOU!

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References


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• 130/5/524 [pii]
References