A Retrospective Review of Timely Intervention for Children with Congenital Microtia and Aural Atresia in a Multidisciplinary Clinic

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Background

Microtia and Aural Atresia (birth abnormalities of the ear):

- Prevalence: 0.83 to 17.4 per 10,000 births in varying degrees; unilaterally in 79% to 93% of cases (Luquetti, Heike, Hing, Cunningham, & Cox, 2012; Luquetti, Leoncini, & Mastroiacovo, 2011)
- Children born with unilateral microtia typically have normal hearing sensitivity in the contralateral ear (Kelley & Scholes, 2007). They are at increased risk for delayed speech-language, academic, and social-emotional development and decreased quality of life (Joint Committee on Infant Hearing, 2000; Lieu, 2015).
- A diagnostic hearing evaluation should be completed soon after birth to determine hearing status for both ears (Kelley & Scholes, 2007).
- Patients should be referred to an otolaryngologist to accurately understand embryology, surgical options, address medical concerns related to the diagnosis, and obtain appropriate imaging.

Children's Hospital Colorado Microtia and Atresia Multidisciplinary Clinic:

- Clinic patient population: birth to age 21
- Clinic specialists: pediatric ear, nose and throat specialist and plastic surgeon, anaplastologist, audiologist, and family consultant.
- Created in 2008 to support each child's individual needs and educate caregivers by answering questions concerning microtia, aural atresia, and options for audiologic and reconstructive treatments.

The Joint Committee on Infant Hearing (JCIH) Position Statements:

- Recommend screening infant hearing for defects of the ear ("Joint Committee on Infant Hearing position statement," 1982).
- State that all infants who do not pass a newborn hearing screening receive audiologic evaluation before three months of age ("Joint Committee on Infant Hearing 1994 Position Statement. American Academy of Pediatrics Joint Committee on Infant Hearing," 1995).
- State that infants should receive early intervention (EI) services by six months of age if hearing loss is confirmed (Joint Committee on Infant Hearing, 2000).
- Define permanent bilateral or unilateral, sensory or conductive hearing loss that averaged 30 to 40 decibels or more between 500 Hz to 4000 Hz as targeted hearing loss for universal newborn hearing screening (UNHS) (Joint Committee on Infant Hearing, 2000).

The current literature does not define whether children with microtia and aural atresia receive diagnostic hearing evaluations or meet the evidenced-based JCIH guidelines.

Objective

The intent of this review was to determine whether patients attending the Children's Hospital Colorado Microtia and Atresia Multidisciplinary Clinic received a diagnostic hearing evaluation, early intervention, and if amplification was fit per evidenced-based JCIH guidelines.

Methods

Retrospective chart review was performed on the CHCO electronic medical record system.

Participants:

Patients treated in the CHCO Microtia and Atresia Multidisciplinary Clinic and born between 2012-2017 were included in the review.

- 55 patients met the criteria for inclusion
  - 49 unilateral microtia and aural atresia
  - 6 bilateral microtia and aural atresia

Data collection and preparation:

Research Electronic Data Capture (REDCap) electronic data capture tools hosted at University of Colorado Denver (Harris et al., 2009).

Patient variables collected:

- Year of birth and sex
- Unilateral or bilateral microtia and aural atresia
- Year and age of first visit with the CHCO Microtia and Atresia Multidisciplinary Clinic
- UNHS completed and in Colorado or out of state
- Age and type of first hearing evaluation
- Age of referral to EI
- Age amplification was fit

Results

Patient Variables	Total Count	Missing	Min	Max	Mean	Median	St Dev
Age of Diagnosis	49	6	1	46	7.51	2	11.32
Age of Early Intervention Referral	45	10	1	33	7.09	2	9.87
Age of First Microtia Clinic Visit	55	0	1	54	12.71	6	14.78
Age of Amplification Fitting	43	12	1	55	11.93	7	12.58

Figure 1

All "age" values in the data set are in reference to age of the patient in months.

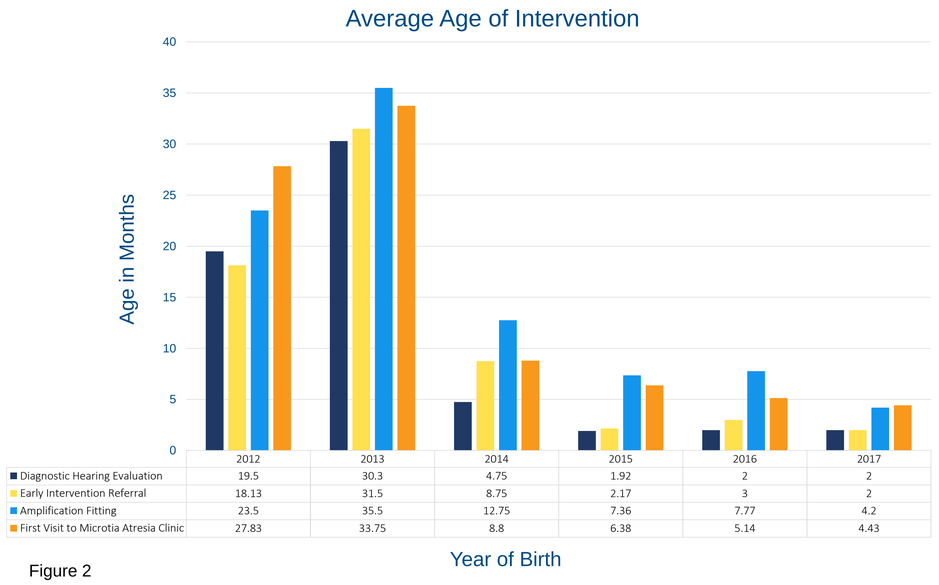

Figure 2

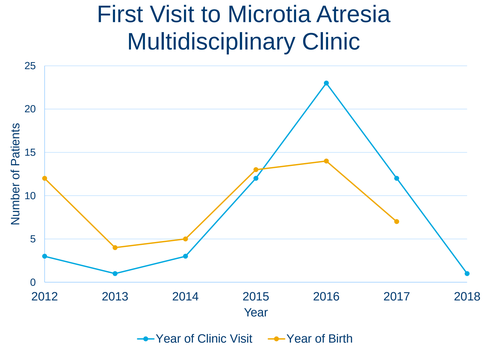

Figure 3

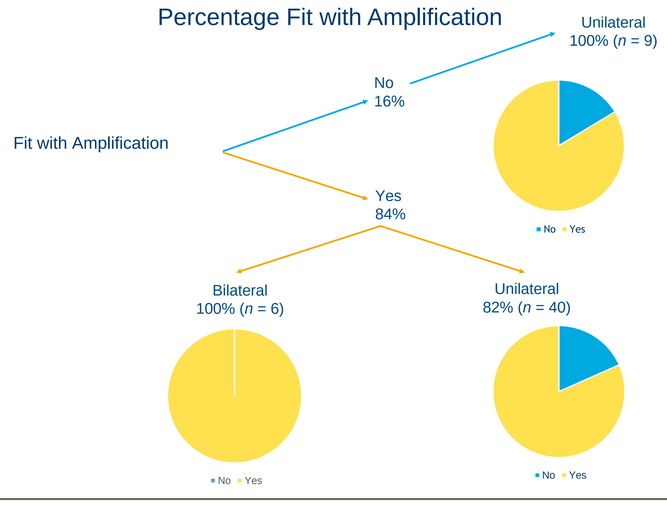

Figure 4

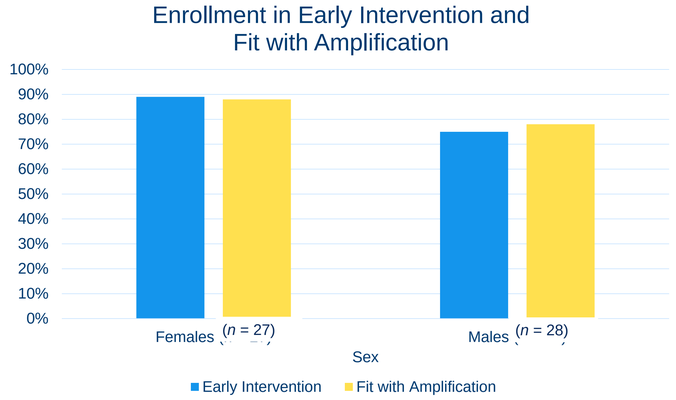

Figure 5

Conclusions

- Over a five-year time span, age of diagnostic hearing evaluation, EI referral, and the fitting of amplification significantly decreased to fit within the current JCIH guidelines (audiologic evaluation before three months of age and EI before six months of age) for children attending the CHCO Microtia and Atresia Multidisciplinary Clinic (Figure 2).
- Patients may not have been born in the same year as their initial visit to the CHCO Microtia and Atresia Multidisciplinary Clinic. The data suggests that over time patients are currently seen in the same calendar year as they were born. This is likely due to an increased referral rate by providers (Figure 3).
- The sooner a diagnostic hearing evaluation is completed, the more likely a patient is referred for EI.
- If a patient with microtia and aural atresia receives EI by 2 months of age or is evaluated through the CHCO Microtia and Atresia Multidisciplinary Clinic by 6 months of age, 90% are fit with amplification.
- The majority of patients (49 out of 55) in this review have unilateral microtia and aural atresia.
- 84% of patients (bilateral and unilateral hearing loss (UHL)) were fit with amplification.
- The majority (82%) of the patients with unilateral microtia and aural atresia were fit with amplification (Figure 4).
- A greater percentage of females (*n* = 27) with microtia and aural atresia than males (*n* = 28) were fit with amplification (Figure 5).
- 43 out of 55 patients received a UNHS in Colorado while; the remaining 12 patients received UNHS in another state (*n* = 5) or did not receive a UNHS (*n* = 7). There was no evidence to suggest that there was a difference in treatment for these populations.

Discussion

- The components that contribute to timely intervention for children with microtia and aural atresia are age of diagnostic hearing evaluation, age of EI referral and evaluation in the CHCO Microtia and Atresia Multidisciplinary Clinic.
- The data suggests that there is an association between the age a patient is fit with amplification and when they initially visit the CHCO Microtia and Atresia Multidisciplinary Clinic. When patients visit the CHCO Microtia and Atresia Multidisciplinary Clinic at a young age, they are fit with amplification sooner. The literature suggests that amplification is recommended for patients with UHL, although it is known that use of amplification is lower in this population when compared to bilateral hearing loss (Fitzpatrick, et al., 2017).
- Specialists in the CHCO Microtia and Atresia Multidisciplinary Clinic counsel patient caregivers to consider amplification at a young age. This may be the reason a majority of the patients with unilateral microtia and aural atresia were fit with amplification. This review has a greater frequency than the literature suggests (7% to 48%) for UHL (Yoshinaga-Itano, DeConde Johnson, Carpenter & Stredler Brown, 2008).
- There is a higher prevalence of unilateral microtia and aural atresia than bilateral cases. This review aligns with current literature (Luquetti, et al., 2011).
- Current literature indicates only 37% to 44% of children with UHL are fit with amplification (Fitzpatrick, Al-Essa, Whittingham & Fitzpatrick 2017; Bagatto et al., 2016). The fit rate in this review is higher.
- According to the literature most patients with UHL have poor compliance with use of amplification. However, they are generally fit at an older age (Yoshinaga-Itano, et al., 2008). Poor compliance with amplification is not specific to this population, but rather children with hearing loss in general based on Walker et al. (2015). Patients in this review were fit with amplification at a young age, therefore, they may have higher compliance.

Limitations

- This is a retrospective chart review. The investigators were unable to control for specific patient variables, therefore, the data may consist of incomplete sets and historic patient charts may not be fully complete.
- The data is biased because the sample of patients only includes patients seen through the CHCO Microtia and Atresia Multidisciplinary Clinic.
- This review does not include newborns born in the last few months of 2017.
- The total of right ear unilateral cases versus left ear was not included in this review.
- A larger sample size would help test the hypothesis that there is no difference in treatment for infants that receive a UNHS in Colorado verses those that do not.

Future Directions

- When patients visit the CHCO Microtia and Atresia Multidisciplinary Clinic at a young age, they are fit with amplification sooner. In order to determine future statistical significance, a larger sample size is needed.
- The data suggests that more females than males are fit with amplification. To determine if a sex bias exists, a larger sample size is needed.
- Reviewing medical records of patients with microtia and aural atresia that are not seen in the CHCO Microtia and Atresia Multidisciplinary clinic will strengthen the ability to determine if patients receive a diagnostic hearing evaluation, EI referral, and the fitting of amplification per the current JCIH guidelines.
- This review found a high percentage of patients were fit with amplification. Future research should monitor long-term use of amplification for children with unilateral microtia and aural atresia

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References on separate page.

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References

Bagatto, M., Moodie, S., Brown, C., Malandrino, A., Richert, F., Clench, D., & Scollie, S. (2016). Prescribing and Verifying Hearing Aids Applying the American Academy of Audiology Pediatric Amplification Guideline: Protocols and Outcomes from the Ontario Infant Hearing Program. *J Am Acad Audiol, 27*(3), 188-203.

Fitzpatrick, E. M., Al-Essa, R. S., Whittingham, J., & Fitzpatrick, J. (2017). Characteristics of children with unilateral hearing loss. *Int J Audiol, 56*(11), 819-828.

Harris, P. A., Taylor, R., Thielke, R., Payne, J., Gonzalez, N., & Conde, J. G. (2009). Research electronic data capture (REDCap)--a metadata-driven methodology and workflow process for providing translational research informatics support. *J Biomed Inform, 42*(2), 377-381.

Joint Committee on Infant Hearing, American Academy of Audiology, American Academy of Pediatrics, American Speech-Language-Hearing Association, Directors of, S., Hearing Programs in State Health & Welfare Agencies (2000). Year 2000 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics, 106*(4), 798-817.

Joint Committee on Infant Hearing 1994 Position Statement. American Academy of Pediatrics Joint Committee on Infant Hearing. (1995). *Pediatrics, 95*(1), 152-156.

Joint Committee on Infant Hearing position statement. (1982). *ASHA, 24*(12), 1017-1018.

Kelley, P. E., & Scholes, M. A. (2007). Microtia and congenital aural atresia. *Otolaryngol Clin North Am, 40*(1), 61-80, vi.

Lieu, J. E. (2015). Management of Children with Unilateral Hearing Loss. *Otolaryngol Clin North Am, 48*(6), 1011-1026.

Luquetti, D. V., Heike, C. L., Hing, A. V., Cunningham, M. L., & Cox, T. C. (2012). Microtia: epidemiology and genetics. *Am J Med Genet A, 158A*(1), 124-139.

Luquetti, D. V., Leoncini, E., & Mastroiacovo, P. (2011). Microtia-anotia: a global review of prevalence rates. *Birth Defects Res A Clin Mol Teratol, 91*(9), 813-822.

Walker, E. A., McCreery, R. W., Spratford, M., Oleson, J. J., Van Buren, J., Bentler, R., . . . Moeller, M. P. (2015). Trends and Predictors of Longitudinal Hearing Aid Use for Children Who Are Hard of Hearing. *Ear Hear, 36 Suppl 1*, 38S-47S.

Yoshinaga-Itano, C., DeConde Johnson, C., Carpenter, K., & Stredler Brown, A. (2008). Outcomes of children with mild bilateral hearing loss and unilateral hearing loss. *Semin Hear, 29*(2), 196-211.