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Original Article

Hearing Aid Uptake in Children with Unilateral Microtia and Canal Atresia: A Comparison between a Tertiary Center and Peripheral Centers

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OBJECTIVES: To review the trialing and uptake of hearing aids in children with unilateral microtia or canal atresia, known collectively as congenital unilateral conductive hearing loss (CUCHL), observed in a tertiary hospital and local peripheral services.

MATERIALS and METHODS: A retrospective review of medical records for patients with CUCHL was conducted using data from a shared audiology database at a tertiary children's hospital.

RESULTS: We identified 45 patients with CUCHL and excluded seven of them due to missing data. Of the 38 patients, 16 (16/38, 42%) did not have any subjective hearing complaints. Furthermore, 32% (12/38) of patients attended audiology at a tertiary centre and 83% (10/12) from this group trialled a hearing aid. In comparison, 46% (12/46) whose audiology care was delivered peripherally trialled aiding. Of the patients from the tertiary center, 58% (7/12) are still using a hearing aid compared to 27% (7/26) of patients from peripheral centers.

CONCLUSION: Our analysis shows that patients with CUCHL are more likely to try hearing aids and continue using them if their audiology care is in a tertiary center. Allowing for a small sample size, this may indicate a health inequality. Agreeing on minimum standards for the management of patients with CUCHL or managing them in a designated center could increase consistency.

KEYWORDS: Microtia, bone conduction hearing aids, conductive hearing loss, osseointegration

INTRODUCTION

Microtia is a congenital malformation of the auricle ranging from minimal abnormalities to anotia, which is the complete absence of the external ear [1]. Microtia is combined with atresia or stenosis in up to 90% of patients, and there is a high correlation between the degree of microtia and the frequency of external and middle ear abnormalities [1-3]. Estimates suggest that 80-90% of patients with microtia and canal atresia have moderate-to-severe conductive hearing loss on the affected side [1, 3]. Microtia has a proclivity to affect the right side and is at least 2.5 times more common in males than in females. Most cases of microtia are sporadic and isolated, but microtia can occur as part of a syndrome, examples of which include the oculo-auriculo-vertebral syndrome (OAVS), Down syndrome, and Treacher Collins syndrome [1, 3].

Children with bilateral canal atresia typically have a moderately severe conductive hearing loss, which mandates early aiding. Despite normal hearing on the contralateral side, unilateral microtia and canal atresia are linked to speech and language delay and learning disabilities, due to the varying degrees of conductive hearing loss [4, 5]. Evidence suggests that children with microtia suffer from poor academic performance and behavioral problems with one study even suggesting a low intelligence quotient [4-6].

Moreover, recent evidence has shown an increased risk of middle ear pathology in children with microtia, particularly the syndromic subgroup where craniofacial abnormalities are inherent [1]. With limited surgical options for restoring binaural hearing to a

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satisfactory level, early hearing rehabilitation with bone conduction hearing aids has been suggested [2].

The goal of our study is to elucidate the uptake and continued use of hearing devices in children with CUCHL in our region and identify differences in service provision between a tertiary hospital and peripheral centers.

MATERIALS AND METHODS

A retrospective review of patients with microtia and canal atresia was conducted using data from a shared audiology database at a tertiary children's hospital between 1996 and 2015.

The study was performed as part of a service evaluation project, and therefore formal ethical approval was not required.

Our study was a retrospective database review, and as such informed consent was not sought. Furthermore, the data used in our work are anonymized, and there is no identifiable patient information.

RESULTS

We identified 45 patients with CUCHL aged 0-19 years. Seven patients were excluded due to missing data. Figure 1 illustrates the sex distribution of the study group. In the remaining 38 patients, OAVS was the most common syndromic association (32%), with the majority being isolated cases.

Sixteen patients (42%) did not have any subjective hearing complaints, and only two from this subset (13%) were offered a hearing aid trial. Across the region, 22 patients (58%) trialed an amplification device, and 64% (14/22) patients found it useful and are still using the device (Figure 2). Furthermore, 32% (12/38) of patients attended audiology at a tertiary centre and 83% (10/12) from this group trialled a hearing aid. In comparison, 46% (12/46) whose audiology care was delivered peripherally trialled aiding. Of the patients from the tertiary center, 58% (7/12) are still using a hearing aid compared to 27% (7/26) of patients from peripheral centers.

There were no significant differences in the severity of hearing loss between patients whose care was provided at the tertiary center and those whose were not; however, it could not be concluded that the tertiary center only looked after patients with severest forms of hearing loss. Further, there was no correlation between the severity of hearing loss and hearing aid uptake or refusal.

A bone conducting hearing aid (BCHA) on a soft band was better tolerated than a contact mini device; all patients who trialed and stopped using an amplification device had used a contact mini device, which they had found either uncomfortable or unhelpful (Figure 3).

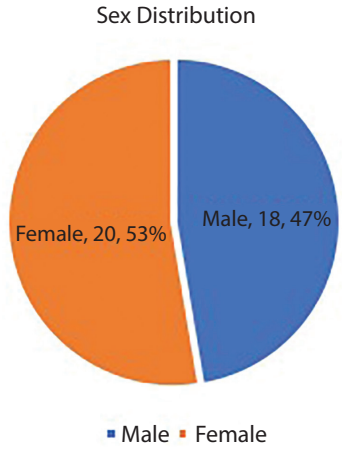

Figure 1. Sex distribution.

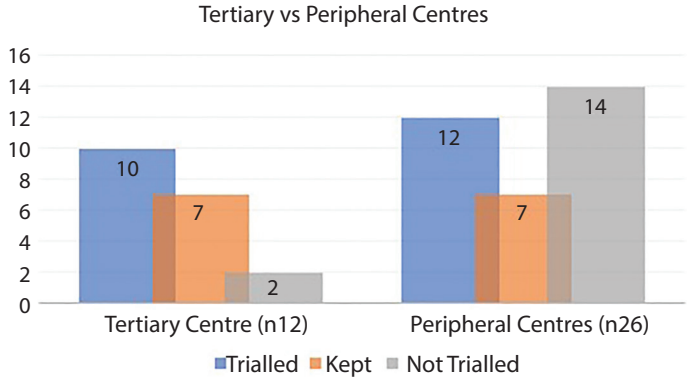

Figure 2. Tertiary versus peripheral centers.

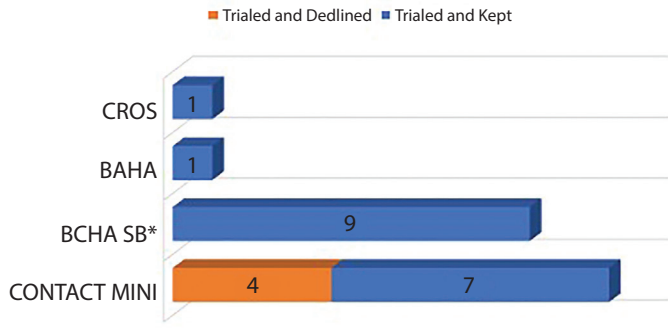

Figure 3. Devices trialed.

MAIN POINTS

- Children with unilateral microtia are at risk of speech and language delay and learning disabilities despite normal hearing on the contralateral ear.
- Binaural hearing is critical to child development and should be restored as early as possible.
- Management of children with microtia requires a multidisciplinary approach (MDT) with a particular focus on parental education, early hearing assessment and rehabilitation.
- Tertiary centres maybe more equipped to address the complex audiological needs of children with microtia.

DISCUSSION

This study retrospectively evaluated the offer, uptake, and continued use of hearing devices in children with CUCHL. We explored the variations in service provision across our region. Until recently, there has been a lack of studies investigating the effects of unilateral conductive or sensorineural hearing loss on educational and social development. Emerging research from audiology and neurosciences illustrates the advantages of binaural hearing over unilateral hearing through binaural summation, squelch, and the head shadow effect [7,8].

Binaural summation is predicated on the brain perceiving sound presented to both ears as louder than sound only presented to one ear despite similarities in sound intensity. Binaural squelch implies the ability of the brain to segregate extraneous background noise to improve sound clarity. The head shadow effect is the phenomenon found in the case of unilateral hearing loss (UHL), where sound arising from the deaf side must travel around the head to the better hearing ear, thus diminishing sound clarity and the ability to localize sound accurately. These three principles, in concert, highlight the potential difficulties caused by UHL despite normal contralateral hearing in one ear-the higher order processing in the brain is lost [7,8]. The corollary is to aid patients with CUCHL, thereby ensuring the highest utilization of higher order brain function.

Our study found that patients with CUCHL are more likely to try hearing aids if their audiology care is administered in a tertiary center (83% versus 46%). Further, 58% (7/12) from the tertiary center are still using a hearing aid compared to 27% (7/26) of patients from peripheral centers. The reasons for this are not obvious, but we hypothesize that variations in resources and practices across our region might be contributory.

At the tertiary center, all children with CUCHL are seen in a dedicated multidisciplinary team (MDT) setting, and potential implications of unilateral conductive hearing loss are discussed with the parents considering the children's choices. Specific issues such as self-esteem and stigma, which are common in patients with CUCHL [9] and recognized as potential barriers to hearing aid uptake, are tactfully addressed in the MDT. A BCHA on a soft band is offered at the earliest opportunity, and patients have regular audiology reviews where hearing tests are conducted and potential issues addressed early. This type of service and multi-professional involvement is not uniformly available across the studied region, which might explain the variation in the service offered to patients.

The other putative explanation is rooted in the traditionally held belief that children with CUCHL attain normal developmental milestones like their peers with binaural hearing. As discussed above, this notion has only recently been challenged in the literature, and as such, practices across the different units in our region might still be evolving toward a more proactive approach to restoring binaural hearing. It is likely that parents' decisions are influenced by the suggestions made by the healthcare practitioner they access: if the healthcare practitioner attaches less significance to the potential problems associated with a lack of binaural hearing, they are unlikely to give a strong recommendation in favor of trying hearing aids. This inevitably affects hearing aid uptake.

The impact of binaural stimuli on long-term plasticity is significant in the context of CUCHL. There is evidence that children as young as 5 years of age with normal hearing have binaural processing capabilities comparable to adults [10]. With limited evidence for acquiring binaural processing skills in patients with CUCHL given an amplification device as adults [11], some authors have suggested aiding at the earliest opportunity, regardless of speech and language, in order to fully harness the benefits of binaural hearing [12].

Unfortunately, data on other markers of hearing difficulties such as challenging behavior and poor educational and language acquisition were not readily available for our study. This would have been useful with regard to analyzing the impact of CUCHL on children who declined hearing aids and on those who are still using one. Nevertheless, we posit that patients with CUCHL should be alerted to the possible benefits that binaural hearing confers regarding improved sound perception amidst background noise, better sound localization, and reduced head shadow effect. Uptake of hearing aids at the earliest opportunity could help prevent some of the adverse outcomes associated with CUCHL. Clinicians should discuss these issues in detail, going beyond simple enquiries as to whether a patient has any hearing concerns. A trial of an appropriate hearing aid should be offered.

A further observation from our study is that at least 64% of patients who trialed a hearing aid found the devices useful. A BCHA on a soft band was better tolerated than a contact mini device, with 64% (7/11) of patients stopping use beyond the trial period due to discomfort (71%; 5/7) or poor hearing outcomes. Eight percent (2/11) of patients who switched from a contact mini device to a BCHA on a soft band found the latter useful. The majority of patients still using an amplification device are using a BCHA on a soft band. In addition, 32% (7/22) of patients who trialed but are no longer using amplification devices had trialed a contact mini device, which they either found unhelpful or painful. It is unclear whether all patients who have discontinued use were offered a different hearing aid. With some of the patients who could not tolerate a contact mini finding a BCHA on a soft band useful, it could be argued that those who stopped using a device after only trialing a contact mini could have found a BCHA on a soft band useful if offered. This highlights the importance of tailoring device options to individual patient needs.

Numerous studies on osseointegrated devices (OIDs) in patients with CUCHL have demonstrated good results with regard to sound perception in background noise, speech recognition, and quality of life [12-14]. A systematic review showed that OIDs confer enormous hearing benefits in patients with congenital atresia and are superior to canalplasty [15]. In our study, only one patient has had an OID, and unfortunately, there was insufficient data to identify the reasons behind this. This is made more challenging by the fact that our patients are examined at multiple sites, though surgery is only performed at the tertiary center. This system relies on practitioners working at other centers in the region referring to the tertiary center. Furthermore, age is a restrictive factor in performing OID surgery for children, and in our unit, it is only offered to children older than 5 years. Using a BCHA on a soft band tends to be a clinical precursor to using an OID. It is reasonable to assume that some of the patients in this study will be offered an OID as they get older, considering the relatively high number already using a BCHA on a soft band.

Limitations

The retrospective nature of our study leaves it prone to the inherent limitations of such a design. Some patients were excluded due to missing information, thus making their data unusable. The small sample size afforded no significant statistical analysis, and these results may not be generalizable. However, it is opined that some of the themes arising from this study, such as global outcome measures for patients who are aided early despite normal hearing on the contralateral side and those that are not, deserve further exploration using a more robust research methodology, which might involve a national research collaborative.

CONCLUSION

Despite the small cohort of patients, the differences highlighted in this study may suggest health inequalities between patients managed in a tertiary center and those seen in peripheral centers. Patients with CUCHL are more likely to try hearing aids and continue using them if their audiology care is in a tertiary center. We propose a robust MDT-based approach to the management of patients with CUCHL, focusing on early assessment and intervention. We further suggest a regional agreement on evidence-based minimum standards for the management of this patient group to standardize care and increase consistency. Alternatively, patients with CUCHL should be managed in a dedicated unit that is appropriately resourced to manage their needs, which vary in complexity.

Ethics Committee Approval: The study was performed as part of a service evaluation project and therefore formal ethical approval was not required.

Informed Consent: Informed consent is not necessary due to the retrospective nature of this study.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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