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The role of bone conduction hearing aids in congenital unilateral and bilateral conductive hearing loss

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Introduction

Amongst the spectrum of conditions encountered by Otolaryngologists is congenital unilateral and bilateral conductive hearing loss. Causes of congenital unilateral and bilateral conductive hearing loss include, but are not limited to, ossicular malformation, stapes fixation, and external auditory canal (EAC) atresia with or without microtia (1,2). These conditions are not mutually exclusive and often occur together (3,4).

The optimal approach for hearing rehabilitation in children with congenital unilateral and bilateral conductive hearing loss is unclear. Management options for EAC atresia include reconstructive surgery, observation, or use of a bone conduction hearing aid, or some combination thereof (3). EAC atresia repair is an option in select cases; however, not all parents elect to proceed with surgery, and not all atresia cases are amenable to surgical repair. For purposes of this review we will focus on the role of bone conduction aids for hearing restoration in this patient population. For more information on EAC atresia repair for hearing restoration, the reader is directed to Service and Roberson (5).

Bone conduction hearing aids can be worn on a fabric headband, referred to as a soft band, or attached to an implant that is surgically affixed to the skull, referred to as a bone-anchored hearing aid (BAHA). A BAHA consists of a titanium fixture, which is osseointegrated into the skull, a sound processor that converts sound waves into vibrations, and an abutment that transmits these vibrations to the implanted fixture (6). This system allows for sound to be transmitted directly to the cochlea through the skull, thereby circumventing any external or middle ear anomaly or pathology (7). With a soft band, the sound processor is mounted on a fabric headband and the signal is transcutaneously conducted through the skull to the inner ear. Soft bands are suitable for children who are too young to receive the BAHA implant or as a trial in patients who are contemplating BAHA implantation.

EAC atresia is one of the most common indications for BAHA implantation in children (8-10). However, traditionally children with congenital unilateral conductive hearing loss (UCHL) secondary to EAC atresia are not offered hearing amplification (11,12). Instead, when the child approaches school age, those identified to have language,
social, or academic concerns will be offered a soft band trial. Those who benefit from this trial are then considered for BAHA implantation. This approach is informed by the traditional thinking that a UCHL does not result in any detrimental effects on language and social development (11,13).

In the case of congenital bilateral conductive hearing loss (BCHL), the conventional approach is to offer a soft band with a single processor, proceeding to a unilateral BAHA when technically feasible. Bilateral BAHAs have traditionally not been thought to offer any additional benefit because of the small attenuation of sound when transmitted via bone conduction (14). Theoretically, one BAHA should also stimulate the cochlea on the opposite side and therefore, binaural hearing may be achieved with a unilateral BAHA. It has been reported in the adult population, however, that bilateral BAHA implantation does incur additional benefit compared with unilateral implantation (15,16). Whether this benefit is also seen in children is unclear.

This review examines the recent literature on the use of bone conduction hearing aids in pediatric patients with congenital UCHL and BCHL secondary to EAC atresia. A thorough search of Medline was completed using the MeSH terms “bone-anchored hearing aid, soft band, unilateral hearing loss, and bilateral hearing loss”. Studies that did not involve a pediatric population (age <18 years) or did not include audiological measurements as the primary outcome were excluded, with the exception of quality of life studies, which were thought to supplement studies with more objective outcomes. We also excluded studies that examined children with sensorineural hearing loss.

**Bone Conduction Hearing Aids in Congenital Unilateral Conductive Hearing Loss**

The traditional approach to congenital UCHL wherein no conduction aids are provided until there is evidence of issues in school or social settings is based on the assumption that a UCHL does not have any detrimental effects on language or social development. A systematic review performed by Cho Lieu in 2004, however, concluded that there does indeed appear to be an association between unilateral hearing loss and an increased incidence of developmental and educational concerns, including grade failures and behavioural issues (17). A few possible risk factors were proposed, including lower cognitive ability, right-sided hearing loss, and severe-to-profound hearing loss; however,
any definitive relationships could not be determined. These findings suggest that there may be a role for earlier intervention in children with congenital UCHL, prior to the onset of any developmental or academic issues.

Saliba et al. (2010) prospectively investigated 17 children with congenital conductive hearing loss, 14 of which were unilateral (18). Pure tone average thresholds and speech discrimination were measured prior to and following soft band application, as well as following BAHA implantation. They found a significant gain in pure tone thresholds following BAHA implantation compared with the soft band. This emphasizes the importance of osseointegration in maximizing hearing gain. The average gain in speech discrimination with a BAHA was found to be 22.1%, with the greatest gain occurring when speech and noise were separated by 90°. In other words, speech understanding was improved in the setting of background noise. This underscores the utility of a BAHA in the classroom, where children are faced with the task of hearing and understanding speech in a noisy environment.

Kunst (2008) performed a prospective cross-sectional study on 11 children with congenital UCHL (19). Audiological testing was performed an average of 35 weeks following BAHA implantation. They did not find a significant improvement in sound localization. This was in contrast to patients with acquired UCHL, whereby studies have found a significant improvement in sound localization using similar testing conditions (20). For speech discrimination, however, Kunst found that there was significant improvement in 63% of the patients for whom data was available. Similar to Saliba, the authors concluded that the BAHA is beneficial for speech discrimination in noisy environments where speech and noise are spatially separated.

Priwin (2007) prospectively investigated 6 children with congenital UCHL, with 4 undergoing BAHA implantation (21). Similar to Kunst, no benefit was found for sound localization. However, there was an improvement in speech discrimination at a signal-to-noise ratio of 0. This represented the most difficult condition, where the level of noise equaled that of the speech signal.

The one consistent finding among the Saliba, Kunst and Priwin studies is that speech discrimination was improved with a BAHA, particularly in noisy environments where speech and noise were spatially separated. The classroom is a setting that closely
resembles this speech and noise arrangement. The extrapolation of these findings is that BAHAs can be particularly helpful in school. This corresponds to studies that have reported high compliance with BAHA use, particularly in the school setting (12,22).

While audiometric evaluations are important ways of objectively determining the outcome of an intervention, the quality of life measures in children with congenital UCHL who are fitted with a BAHA must also be considered, as not all benefit can be measured quantitatively. De Wolf (2011) administered quality of life questionnaires to 15 children with congenital UCHL following BAHA implantation (22). One hundred percent of children used their BAHAs on a daily basis. Forty-seven percent used it for greater than 8 hours a day, while 40% used it between 4 to 8 hours; the remainder used it between 2-4 hours. Sixty-seven percent believed that the benefit from the BAHA outweighed the process and risks associated with undergoing the procedure. Health-related quality of life was assessed using the Glasgow Children’s Benefit Inventory (GCBI). The greatest benefit was seen in the domain of learning. This correlates with another study looking at the same quality of life measure, where learning was also found to have the greatest improvement following BAHA implantation in children with congenital UCHL (12).

The quality of life data for BAHAs in children with congenital UCHL show a benefit in the realm of learning. This supports the audiological findings discussed earlier, in which children gain the most benefit from a BAHA in noisy situations such as the classroom.

**Bilateral Bone Conduction Hearing Aids in Congenital Bilateral Conductive Hearing Loss**

Children with congenital BCHL are typically fitted with a soft band with one processor as soon as possible, followed by the implantation of one BAHA. One BAHA is believed to be sufficient because of the traditional thought that the small attenuation of vibrations in the skull would allow for the contralateral cochlea to be stimulated (23). However, transcranial attenuation of sound via bone conduction can range from -5 and +20 dB between 250-10,000 Hz depending on skull thickness, which is highly variable.
among patients (24,25); therefore, the contralateral cochlea cannot be assumed to be receiving the same signal intensity as the side which receives the stimulus.

Our search yielded only one prospective cross-sectional study that investigated the benefit of bilateral BAHAs in children with congenital BCHL. Priwin (2007) performed audiological evaluations in 9 children with BCHL, 7 of whom had a congenital etiology stemming from EAC atresia (21). Three children underwent bilateral BAHA implantation and 6 underwent unilateral BAHA implantation. A significant improvement was found with sound field pure tone average testing following unilateral BAHA implantation, with no additional gain with a second BAHA. Similarly, there was no gain found in speech discrimination following a second BAHA. They did find, however, that sound localization improved with the addition of a second BAHA.

In a retrospective review of 27 children with BCHL, with 18 cases being congenital in etiology, Dun (2010) assessed the quality of life of children fitted with bilateral BAHAs based on the Glasgow Children’s Benefit Inventory (26). Overall, there was a benefit of +38 (where -100 indicated maximum deterioration and +100 indicated maximum benefit). A gain was seen across all domains, with the greatest benefit seen in the domain of learning.

There is limited evidence on bilateral BAHA implantation in children with congenital BCHL. Existing studies suggest that bilateral BAHAs may confer an improvement in sound localization as well as an improvement in quality of life, especially for learning.

**Discussion**

While the benefit of a BAHA in congenital UCHL is not consistent across audiological measures, there does appear to be benefit in speech discrimination. The quality of life studies showed that there are children with congenital UCHL who benefit subjectively from BAHA implantation, particularly with regards to learning. Furthermore, there is enough evidence to raise concern that unaided unilateral hearing loss may have negative effects on development. Finally, there is very little morbidity associated with wearing a soft band. To the best of our knowledge, there have never been any reported complications stemming from soft band use. While the audiological
outcomes are not as robust as compared to an osseointegrated implant, it would seem that the soft band is a preferred alternative to no amplification. Given these factors, it may be reasonable to fit every newborn with congenital UCHL with a soft band, with the option for a BAHA, or even EAC atresia repair, later in life.

Similarly, there is only preliminary data on bilateral BAHA implantation in children with congenital BCHL. Theoretically, bilateral implantation is beneficial as it facilitates binaural hearing. Studies thus far suggest an improvement in sound localization as well as quality of life with bilateral BAHA implantation. An additional processor on the soft band carries minimal morbidity; therefore, we believe it is reasonable to consider every child with congenital BCHL for a bilateral soft band trial as there is a possibility for additional benefit compared to a unilateral soft band. More definitive intervention, such as unilateral or bilateral BAHA implantation, or potentially EAC atresia repair, can then be entertained when older.

Conclusion

The optimal management approach for hearing rehabilitation in children with congenital unilateral and bilateral conductive hearing loss is currently unclear. More prospective studies with larger sample sizes are needed to better elucidate the possible benefits of BAHA use in these unique populations. In the mean time, it would be reasonable to trial unilateral soft bands in children with congenital UCHL and bilateral soft bands in children with congenital BCHL. The rationale is that there is the likelihood for benefit in some children, as well as the absence of any significant risks associated with this intervention. Should the soft band trial prove to be beneficial for the individual child, subjectively or objectively, then a BAHA (or bilateral BAHA) could be considered, or when and if appropriate, EAC atresia surgery undertaken.
References

12. Kunst SJW, Hol MKS, Mylanus EAM, Leijendeckers JM, Snik AFM, Cremers CWRJ. Subjective benefit after BAHA system application in patients with


22. de Wolf MJF, Hol MKS, Mylanus EAM, Snik AFM, Cremers CWRJ. Benefit and quality of life after bone-anchored hearing aid fitting in children with unilateral or


