Effectiveness of Bone Conduction Hearing Aids in Young Children with Congenital Aural Atresia and Microtia

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Background: The aim of this study was to assess the effectiveness of bone conduction hearing aids in children under 2 years old who have congenital microtia and atresia.

Material/Methods: This prospective study involved 42 children under 2 years old with congenital microtia and atresia who were divided into 2 groups: 21 with unilateral defect and 21 with bilateral defect. All children were provided with bone conduction hearing aids on a softband. Air and bone auditory thresholds were assessed by auditory brainstem responses (ABRs). The LittlEARS questionnaire was used to evaluate auditory development at baseline and after 6 months of hearing aids use. Behavioral observation audiometry (BOA) was used to assess auditory thresholds and compare aided and unaided hearing.

Results: After 6 months of hearing aid use, the total score of the LittlEARS questionnaire in children with unilateral defect was 24±5.60, while children with bilateral defect achieved a result of 26.29±6.17. Hearing thresholds in both groups with bone conduction hearing aids improved significantly and approached the normal level.

Conclusions: Our results confirm that bone conduction hearing aids provide an effective method of auditory rehabilitation for children with conductive and mixed hearing loss caused by microtia and atresia. Using bone conduction hearing aids in such children is crucial for proper hearing, speech, and language development.

Keywords: Bone Conduction • Hearing Aids • Hearing Loss

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Background

Early use of a prosthesis and auditory rehabilitation allow the auditory development of children with hearing loss to proceed normally [1]. There are various options for using hearing aids based on either air or bone conduction of sound. For very young children with congenital microtia and aural atresia, the first and often the only form of auditory stimulation available is the use of bone conduction. Although there are many studies assessing the use of various implantable devices and conventional hearing aids among children in Poland and other countries, there are very few studies evaluating the use of bone conduction hearing aids in this age group. Children with microtia and atresia of the external auditory canal are a unique group. The low number of published papers may be due to the low frequency with which microtia and atresia appear and the difficulties associated with assessing the benefits of such aids in young children. Reliable assessment of the effectiveness of bone conduction devices in children under age 24 months requires the use of appropriate research and assessment tools [2].

Hearing impairment is one of the most common defects found at birth [1]. Microtia is a congenital deformity affecting the outer ear (ranging from it not being fully developed, to its complete absence). The reported prevalence varies among geographic regions, from 0.83 to 17.4 per 10 000 births. The condition occurs more frequently in males (60-80%) and is usually unilateral (77-93%), with the right ear being affected in approximately 60% of cases. Microtia is usually accompanied by atresia, a condition in which the external auditory canal is absent or closed [3]. The sooner a hearing defect is detected, the sooner therapy and rehabilitation can begin, opening the door to proper development and regular functioning. The time at which a child is provided with a hearing aid or other assisted hearing device is crucial to the child’s development. According to guidelines on the selection of hearing aids, a child with hearing impairment should be diagnosed before 3 months of age and fitted with a hearing aid before 6 months [4-6]. The selection and fitting of hearing aids for very young patients is a complex process that requires the cooperation of specialists in various areas. Detailed diagnostics, appropriate for the patient’s age, are carried out prior to providing hearing aids, allowing assessment of each ear separately [6]. Awareness by parents and specialists about the solutions available and the possibilities of using specific hearing aids is particularly important. The most important element in the selection of hearing aids is proper fitting and the ability to assess the device’s effectiveness [7]. In very young patients, observation of the child’s reactions to audiological testing is the only indication available, allowing only a preliminary assessment of the device's benefits. The child alone cannot communicate that the hearing aid is correctly fitted and working properly. Monitoring auditory rehabilitation after fitting a hearing aid allows the child’s progress in developing hearing skills to be continuously assessed [8,9]. Lack of progress or a delay in auditory development may indicate inappropriate fitting of the hearing aid or an incorrect course of auditory rehabilitation.

Children under the age of 2 years who have been diagnosed with conductive or mixed hearing loss due to external and/or middle ear defects are a challenging group of patients. For this constantly expanding group of youngest patients it is extremely important to be able to assess the effectiveness of the hearing prostheses used. This assessment cannot be based solely on examinations such as a behavioral hearing test; audiological diagnosis needs to be supplemented with appropriately prepared and validated surveys [8,9]. A tool that has been translated and validated into Polish language for assessing auditory development and the effectiveness of medical intervention used for children under 24 months of age is the LittleEARS questionnaire [9]. The questionnaire has been validated and translated into 26 languages. The questionnaire consists of 35 questions assessing the child’s individual hearing skills, including the identification, discrimination, or location of a sound source (Appendix). The results of the survey allow the current auditory age of the child and the delay in auditory development (relative to correctly hearing children) to be determined.

Currently, there are many bone conduction devices (BCDs) available for hearing rehabilitation. The devices are divided into implantable and non-implantable. BCDs that vibrate bone through the skin are referred to as skin-driving devices; this group includes bone conduction hearing aids that are attached with softbands, frames of glasses, or adhesive adapter. The devices differ in design, mounting, and fit range, but the operating principle is the same. The audio processor picks up sound waves, converts them into vibrations, and transfers them to the mastoid bone directly to the inner ear (bypassing the outer and the middle ear). Through bone conduction, the sound information reaches the inner ear and can help people with conductive and mixed hearing loss (CHL, MHL) and single-sided deafness (SSD). Bone conduction hearing aids mounted on bands are recommended for patients whose bone conduction threshold does not exceed 45 dB HL, or 20 dB HL in the case of devices mounted with a patch. They can be used from the first months of a child’s life, which is an important advantage of the device. The most likely candidates for bone conduction hearing aids are children or adults who have severe outer or middle ear malformations, or those with unilateral hearing loss. This type of hearing solution is also recommended in cases of chronic ear infections and allergies to conventional air conduction (AC) hearing aids. The use of bone conduction hearing aids on a softband is a valuable first method of hearing rehabilitation in children who are too young for implantation [10-22].
Bone conduction hearing aids are a widely known and used solution; however, few medical facilities conduct research on how effective they are in use. There are currently very few reports in the literature on the assessment of auditory development of children under age 2 years who use bone conduction hearing aids.

An important element of this work was to outline available modern options for auditory prostheses in children, with particular emphasis on bone conduction of sound. The age of the child and the degree and place of hearing damage were considered important factors. In young children, for whom it is too early to implant due to their age and anatomical conditions, bone conduction hearing aids should be used as soon as possible to stimulate hearing.

The main objective of this study was to assess the effectiveness of bone conduction hearing aids in children under 2 years of age who had microtia and atresia. The results were based on audiometric tests and a questionnaire.

The specific objectives were:
1. Assessment of the effectiveness and compensation of conductive and mixed hearing loss using non-implantable bone conduction hearing aids in children up to 2 years of age with unilateral or bilateral hearing loss based on audiometric tests and the LittlEARS questionnaire.
2. Determine the delay (if present) in auditory development in children with bilateral and unilateral hearing loss with bone conduction hearing aids (compared to children with normal hearing).
3. Comparison of the effects of bone conduction hearing aids in children with unilateral hearing loss with children with bilateral hearing loss.

Material and Methods

Studies on the effectiveness of bone conduction hearing aids in young children are rare. In global publications, research groups usually do not exceed 20 people. In this study, the research group consisted of 42 children with one- or two-sided microtia and atresia. The results were based on audiometric tests and a questionnaire.

The research process was divided into 2 stages. A combination of behavioral and physiological measures was used to assess children’s auditory development. During the first visit to the Institute of Physiology and Pathology of Hearing, all children were examined for auditory brainstem responses (ABRs). Among the various procedures related to the diagnosis of hearing loss in infants from birth to 6 months of age, the most important and reliable technique is ABR (Joint Committee on Infant Hearing [JCIH] 2007). The study involved both air and bone conduction of sound. All children were tested using the Vivosonic Integrity V500 instrument. Children underwent tone burst auditory brainstem response measurements at 4 selected frequencies: 500, 1000, 2000, and 4000 Hz. Due to anatomical conditions, headphones were used in all children. A complementary study to assess hearing thresholds used behavioral observational audiometry (BOA) over the frequency range 250-6000 Hz. Hearing tests were conducted in a soundproof room. Sounds of varying intensity were presented to the child via calibrated speakers from 1 meter. Sounds consisted of speech or music, of a certain frequency (from 250 to 6000 Hz) and intensity.

Based on the above tests, children were selected and fitted with appropriate hearing aids using bone conduction of sound. All devices were mounted on softbands, and all children used the same bone conduction hearing aids. The patients were selected according to the following criteria: patients under 2 years of age, unilateral or bilateral microtia with atresia, reliable cooperation in audiological tests, hearing thresholds had to comply with the manufacturer’s criteria (less than or equal to 45 dB HL), and the parents needed to have realistic expectations of benefits and to know the limitations of bone conduction hearing aids. Unfortunately, fitting protocols for bone conduction hearing aids for children are still not well developed. In our work, the settings of the devices were based on the results of the ABR test and tests carried out in a free acoustic field with aided conditions. Appropriate software, manufacturer’s recommendations, and a method dedicated to children – DSL (Desired Sensation Level) – were used. To assess the effectiveness of the hearing aids, behavioral hearing testing was carried out over a wide frequency range (250-6000 Hz) with the hearing aids activated. The LittlEARS questionnaire was completed by the child’s parents and was used to assess the current level of auditory development (auditory age and delay in auditory development). The second visit occurred at about 5 months (~2 weeks) after the first visit. It consisted of audiometric evaluation (a BOA test carried out with the fitted hearing aid) and a survey in which parents again filled out the LittlEARS questionnaire.

The study was conducted using 2 groups: children with unilateral hearing loss (UHL) and children with bilateral hearing loss (BHL). The auditory brainstem response (ABR) was tested in all children during the first visit (before using hearing aids). We used the t test to compare hearing thresholds in both groups (UHL group vs BHL group). Auditory development (auditory age and delay in auditory development) was assessed twice – before and after use of the hearing aids – using the LittlEARS questionnaire. To assess progress in hearing development in both groups, ANOVA was used in a mixed scheme with Bonferroni correction for multiple comparisons. Behavioral observation audiometry (BOA) was performed after using hearing aids.
The \( t \) test was used to determine the difference in benefits for children with UHL and children with BHL. Statistical analysis was performed using IBM SPSS Statistics software (v. 24). A \( P \) value below 0.05 was considered statistically significant.

The research methodology used was in line with methods presented to the Bioethics Committee and its decision of 26 October 2018 (KB number IFPS: 23/2018). Parents or legal guardians signed the consent before proceeding with the study.

Results

Demographics

The research group consisted of 42 children with one- or two-sided microtia and atresia and conductive or mixed hearing loss. The group included 16 females and 26 males aged 5 to 19 months (\( M=14.1, \) SD=4.8). Half of the group (21 people) were children with unilateral hearing loss (UHL), and the others had bilateral hearing loss (BHL). There were 13 children who had a one-sided defect in the right ear, and 8 children had a disorder affecting the left ear. Data regarding their sex and age are given in Table 1.

|                | Children with UHL | Children with BHL |
|----------------|-------------------|-------------------|
| Gender         |                   |                   |
| Male           | 17 (81%)          | 9 (43%)           |
| Female         | 4 (19%)           | 12 (57%)          |
| Age (months) (mean±SD) | 14.4±4.8 | 13.7±4.9 |

Table 1. Demographic data of the 2 study groups.

Results of ABR testing for air (AC) and bone (BC) conduction are shown in Table 2.

|                | Children with UHL | Children with BHL | t-test, p-value |
|----------------|-------------------|-------------------|-----------------|
| AC Right ear   | 69-90; 72.31±10.13 (13 ears) | 50-90; 72.86±9.56 | t=0.16; p=0.874 |
| AC Left ear    | 60-80; 67.50±8.86 (8 ears) | 30-100; 72.86±15.21 | t=0.93; p=0.360 |
| BC Right ear   | 10-40; 20.00±9.13 (13 ears) | 10-40; 20.48±10.24 | t=0.14; p=0.892 |
| BC Left ear    | 10-20; 17.50±4.63 (8 ears) | 10-50; 20.48±12.03 | t=0.68; p=0.506 |

Table 2. ABR test results for air (AC) and bone conduction (BC) testing.

The average BOA score in children with UHL was 28.10 (SD=3.35), in the range 25 to 35 dB. The average BOA result in children with BHL was 36.19 (SD=4.16), in the range 30 to 45 dB. The difference between the groups was statistically significant: \( t=6.96; \) \( P<0.001 \). The average BOA result was significantly higher in children with BHL than in children with UHL.

Table 3. Results of LittIEARS.

|                | Children with UHL | Children with BHL |
|----------------|-------------------|-------------------|
| LittIEARS score |                   |                   |
| Before providing with HA | 0-34; 22.33±9.29 | 0-30; 10.95±8.39 |
| After providing with HA | 16-35; 28.24±5.60 | 14-35; 26.29±6.17 |
| Hearing age     |                   |                   |
| Before providing with HA | 4-19; 11.95±5.18 | 1-18; 5.43±4.86 |
| After providing with HA | 8-24; 17.57±5.24 | 7-24; 15.14±5.88 |

Table 3. Results of LittIEARS.

Looking at interactions, an ANOVA for the LittIEARS result was statistically significant: \( F(1,40)=18.33; \) \( P<0.001; \) \( \eta^2=0.314 \).
The average results after use of hearing aids were similar in both groups (mean difference was 1.95, \( P = 0.305 \), \( \eta^2 = 0.026 \)). In both groups, the average LittlEARS score increased significantly from the pre-treatment period compared to the period after the hearing aids were supplied. A larger increase was observed in the group of children with BHL (the average difference was 15.33, \( P < 0.001 \), \( \eta^2 = 0.708 \)). In the group of children with UHL, the average difference was 5.91; \( P < 0.001 \); \( \eta^2 = 0.264 \).

The effect of ANOVA interaction on auditory age was statistically significant: \( F (1.40) = 7.94; \ P = 0.007; \ \eta^2 = 0.166 \). Children with UHL had a much older auditory age than children with BHL, but only in the period prior to hearing aid use (mean difference between groups was 6.52, \( p < 0.001, \eta^2 = 0.307 \)). The mean auditory age on aided hearing was similar in both groups (mean difference was 2.43; \( P = 0.165; \ \eta^2 = 0.048 \)). The mean auditory age increased significantly in both groups; the result was higher in the group of children with BHL (mean difference 9.71; \( P < 0.001; \ \eta^2 = 0.691 \)) than in the UHL group (mean difference 5.62; \( P < 0.001; \ \eta^2 = 0.428 \)).

The effect of ANOVA interaction on auditory development delay was statistically significant: \( F (1,40) = 8.28; \ P = 0.006; \ \eta^2 = 0.172 \). Children with BHL had a greater delay in auditory development.
than children with UHL, but only before the use of hearing aids (mean difference between groups was 5.76; p<0.001; e²=0.268). The average auditory development delay after hearing aid use was similar in both groups (mean difference was 1.95, P=0.098, e²=0.067). The average auditory development delay was significantly reduced in the group of children with BHL (mean difference was 4.33, P<0.001; e²=0.349), while the hearing delay in the UHL group remained at a similar level between unaided and aided (mean difference 0.62, P=0.579).

Figure 1 presents the difference in points obtained on the LittlEARS questionnaire before and after using bone conduction hearing aids in the group of children with unilateral hearing loss. The blue curve refers to the points obtained in unaided hearing, while the orange curve refers to the points obtained with aided hearing (Figure 1).

Figure 2 presents the difference in points obtained in the LittlEARS questionnaire in unaided and aided hearing in a group of children with bilateral hearing loss. The blue curve refers to the points obtained in unaided hearing, while the orange curve applies to points obtained with aided hearing (Figure 2).

Discussion

Children with congenital bilateral microtia with atresia of the external auditory canals constitute a very specific and unique group of patients. Bone conduction hearing aids are the first step in the process of qualifying for an implantable device. Such aids provide a real option for sound stimulation, opening the way to the world of sounds. Detailed evaluation of the device chosen is necessary for the proper adjustment of the hearing aid and for good rehabilitation. Research in this area is not commonly done, and the research group on which the results of this work are based is one of the largest among similar investigations.

This work and the publications by other authors presented below confirm the effectiveness of the use of bone conduction devices in children of various ages. The results reported are based on audiometric tests and questionnaires.

The effectiveness of using bone conduction devices has been shown in a recent study by Kulasegarah et al of patients aged 5-15 years with microtia and/or atresia and moderate bilateral conductive hearing loss [15]. Implantable devices were used to treat all patients. Before receiving the implants, the children had used traditional bone conduction hearing aids mounted on softbands. Audiometric results were compared from when the children used hearing aids and then after implantation. The benefits of both devices were shown to be comparable – both bone conduction devices and implants improve hearing and speech understanding – and there were no statistically significant differences. That is, both solutions perform satisfactorily [15].

Another important element to be considered is the implementation of early auditory intervention and rehabilitation. These programs provide opportunities for proper auditory and speech development, a result which has been demonstrated by Zhang et al [10], who performed a retrospective analysis of hearing and speech development in young children (12 patients aged 3 months to 6 years) who had bilateral microtia and atresia and who had previously used bone conduction hearing aids fixed with a softband. Before and after comparisons were made based on audiometric tests and the IT-MAIS questionnaire, 6 months after using the devices. The free-field air conduction results of the ABR test averaged 73 dB, and after using a bone conduction hearing aid these thresholds changed significantly, with the mean result now being 21 dB, a 52 dB improvement.

The results of our work also show lower thresholds over a wide frequency range, with the mean decrease in air conduction threshold in the free field being 32 dB. Auditory perception in children with bilateral, congenital microtia with atresia of the auditory external canals definitely improved after the use of bone conduction hearing aids. We believe that the use of bone conduction devices in the youngest patients can ensure relatively normal hearing development and avoid future disturbance of voice communication.

Other work confirming the effectiveness of bone conduction hearing aids in allowing normal auditory and speech development includes a study by Wang et al [23], who assessed the hearing development of 20 children (aged 3-21 months) who had bilateral microtia and atresia after using bone conduction devices mounted on softbands and compared it to children with normal hearing. Visual reinforcement audiometry was performed and the IT-MAIS questionnaire was administered during the first visit and at follow-up visits 3 and 6 months after the devices were implanted. The results showed a significant improvement in hearing and proper auditory development. The use of hearing aids is therefore possible in infants, and this enables very early intervention and auditory stimulation.

Christensen et al compared the functional improvement of various skeletal organs in infants and children with bilateral conductive defects, work done at the Arkansas Children’s Hospital [14]. The devices tested included traditional bone conduction devices, Baha devices mounted on soft elastic headbands, and Baha implantable devices. There were 10 participants who had bilateral conductive hearing loss and used traditional bone prostheses; they then switched to a Baha softband, and finally implantation of a Baha bone conduction implant. To evaluate the gain, tonal audiometry was used at 500,
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1000, 2000, and 4000 Hz. Despite differences in gain level, the results showed improved hearing and benefits from all devices used. According to the authors, bone conduction hearing aids should be used as the first choice in medical intervention in patients with conductive/mixed hearing loss for whom conventional hearing aids cannot be used. In young children, for whom it is too early to implant due to their age and anatomical conditions, bone conduction hearing aids should be used as soon as possible so as to stimulate hearing.

One of the goals of this study was to evaluate the auditory development of children after they had been provided with a bone conduction hearing aid, an approach also used by Fan et al [18] with 16 patients aged 3 months to 6 years with bilateral microtia and atresia of the external auditory canals. The study aimed to compare the auditory development of children using bone conduction devices mounted on bands with children with normal hearing. The level of auditory development based on IT-MAIS questionnaire was rated 3 times: during the first visit and at follow-ups at 6 and 12 months. They found that the auditory development of children equipped with bone conduction devices is equivalent to that of normal-hearing children. This finding is similar to that seen in our work. Children with bone conduction hearing aids achieved normal hearing thresholds. The results of the survey used in our work – the LittleEARS questionnaire – showed that children with bone conduction hearing aids achieved a result within the limits for children with normal hearing.

Another aspect of our work is the validity of using bone conduction hearing aids in patients with unilateral hearing loss. In patients with bilateral hearing loss, bone implants give good satisfaction and benefits. However, assessing the benefits is much more difficult in patients with unilateral defects. According to national and international guidelines for the use of hearing aids, it is appropriate to use bone conduction devices in patients with unilateral hearing loss [24]. Despite normal auditory development (deriving from normal hearing in the contralateral ear), patients often report problems with locating the source of sounds, lack spatial hearing, and – which is the biggest problem and source of discomfort for them – impaired speech understanding in noisy environments. Previous studies and numerous publications on children with unilateral hearing loss have shown great difficulties in everyday functioning. Educational problems, deficits in hearing skills, behavioral disorders, and communication barriers resulting from hearing impairment are the most common factors disrupting the proper development of this group of children. They have a particular impact on a young child, for whom the first years of life are the period of acquiring aural and linguistic skills and improving communication [17,25-28].

The work of Kunst et al [29] demonstrates the effectiveness of bone conduction devices in a group of patients (10 adults and 10 children) with unilateral hearing loss. The use of bone conduction devices resulted in hearing benefits and improved understanding of speech in noise in all tested patients.

Doshi et al assessed improvements in the quality of life in 8 children aged 7-12 years with unilateral deafness who were provided with bone conduction implants [30]. Two questionnaires were used to assess the benefits and quality of life: the Glasgow Children’s Benefit Inventory (GCBI) and the Single-Sided Deafness (SSD) Questionnaire. The average level of satisfaction with the implants was 9/10, indicating the correct operation of the devices and good patient satisfaction. Further work is needed in a larger series of children to see if this benefit is statistically significant.

Conclusions

Our research on the use of bone conduction hearing aids in children with congenital microtia and auricular atresia under 2 years of age has demonstrated that:

1. Hearing aids, when using bone conduction, can effectively compensate for conductive and mixed hearing loss in children up to 2 years of age who have unilateral and bilateral hearing loss. The obtained audiometric results confirm this. After using bone conduction hearing aids, children with hearing impairment (either: single-sided or double-sided) achieve better results, their detection thresholds decrease significantly, and their auditory reactions are much better.

2. The use of hearing aids using bone conduction in children under 2 years of age is an effective form of medical intervention. Bone conduction hearing aids in our study group significantly improved the children’s auditory responses and skills. An appropriate level of auditory development (no delay in development) in children using bone conduction hearing aids confirms their effectiveness.

3. Before using the devices, children with unilateral hearing losses obtained better results than those with bilateral defects. After using the devices, this difference almost disappeared, which means a significant improvement in auditory development in children with bilateral hearing loss.

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Declaration of Figures Authenticity

All figures submitted have been created by the authors, who confirm that the images are original with no duplication and have not been previously published in whole or in part.
Supplementary

Appendix: LittleEARS questionnaire.

https://www.hse.ie/eng/services/list/4/audiology/childrens/usefulresourcesforchildrensaudiology/littleears%20questionnaire.pdf

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