American Journal of Medicine and Medical Sciences 2025, 15(7): 2099-2103
DOI: 10.5923/j.ajmms.20251507.03

The Ponto System in Children with Bilateral Congenital Aural Atresia

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Abstract Restoration of hearing in children with bilateral congenital aural atresia presents considerable challenges due to complex anatomical variations and limited surgical options. This study evaluates the efficacy of bone conduction hearing rehabilitation using the Ponto Softband system in infants and toddlers with bilateral microtia and atresia. A cohort of 40 children aged 3 months to 2 years was assessed over a 24-month period using the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), visual reinforcement audiometry, and speech perception tests. The findings demonstrate significant improvements in auditory thresholds and speech recognition scores following the use of Softband-mounted Ponto devices. Compared to normal-hearing peers, children with microtia-atresia showed an initial delay in auditory development, which decreased progressively with consistent use of the device. The study confirms the importance of early auditory rehabilitation to support language acquisition and cognitive development in this high-risk population. The results advocate for the use of non-surgical bone conduction hearing aids prior to auricular reconstruction as a safe and effective approach to managing bilateral aural atresia in pediatric patients.

Keywords Congenital aural atresia, Microtia, Bone conduction hearing aid, Ponto system, Softband, Pediatric audiology, Auditory rehabilitation, IT-MAIS, Speech development, Non-surgical hearing correction

1. Introduction

Congenital malformations of the ear, particularly bilateral aural atresia accompanied by microtia, represent a significant clinical challenge in pediatric otolaryngology. These anomalies are not only rare—with an estimated incidence of 0.5–0.6 per 10,000 live births—but are often associated with complex structural deformities involving the external and middle ear, leading to conductive hearing loss. The anatomical variability of the temporal bone, the frequent co-occurrence of ossicular malformations, and the risk of complications such as facial nerve injury or canal restenosis complicate the prospects for surgical correction.

In children with bilateral aural atresia, the impairment of auditory input during the critical early years of language development can result in delayed speech acquisition, reduced cognitive performance, and adverse psychological outcomes. The first two years of life are particularly crucial, as auditory stimuli during this period play a foundational role in shaping verbal communication and neurocognitive processing. Therefore, early and effective hearing rehabilitation is essential to prevent long-term developmental deficits [1,2,3,4,5,6].

Traditionally, reconstructive surgery has been the mainstay of treatment, but outcomes are variable, and risks remain significant. In recent years, bone conduction hearing systems, such as the Ponto device mounted on a Softband, have emerged as a promising non-surgical alternative. These devices bypass the malformed external auditory canal and ossicles, transmitting sound directly to the cochlea through skull bone vibration. Early use of such technology has the potential to restore auditory perception, support speech development, and improve social integration without the need for immediate invasive procedures.

This study aims to evaluate the auditory development and speech outcomes in infants and toddlers with bilateral microtia and aural atresia using the Ponto Softband system. By comparing these outcomes to normative data from children with normal hearing, the study provides important insights into the benefits and limitations of early non-surgical auditory rehabilitation in this high-risk pediatric population [7,8,9].

Received: Jun. 8, 2025; Accepted: Jun. 27, 2025; Published: Jul. 3, 2025
Published online at http://journal.sapub.org/ajmms

2. Results

Restoring auditory function in children with ear malformations is a complex task that requires the joint efforts of audiologists, otorhinolaryngologic surgeons, and surdopedagogues. Ear malformations constitute 50% of all congenital anomalies encountered in ENT practice. Hearing pathology associated with congenital middle ear anomalies is diagnosed in 1–6% of the population, and ENT anomalies

account for 22% of all developmental abnormalities.

In cases of congenital aural atresia, the anatomical variability of the temporal bone structure, unsatisfactory outcomes from surgical treatment, and high risk of complications necessitate the development of precise indications, contraindications, new treatment strategies, preoperative preparations, and post-treatment rehabilitation protocols. Congenital ear malformations are not common, with a population frequency of about 0.5–0.6 per 10,000 people. However, these statistics do not account for individual symptomatic contributions [10,11,12,13,14].

Various classifications have been developed to determine the surgical strategy and feasibility in ear developmental anomalies. Most are based on computed tomography results of the temporal bone and are intended to predict surgical outcomes.

Among the anomalies of the auditory system, external auditory canal atresia and the associated complete absence or malformation of the ossicular chain, as well as bony adhesions of the middle and inner ear, are frequent. Inner ear anomalies are found in 20% of patients with congenital sensorineural hearing loss.

Numerous studies have explored histopathology and surgical outcomes in isolated congenital ossicular malformations without external ear anomalies. However, there is no universal surgical strategy for isolated middle ear anomalies in the global literature, and most authors recommend various types of ossiculoplasty.

Until recently, many inner ear anomalies were considered absolute contraindications to cochlear implantation due to poor visibility of the tympanic cavity structures during retrotympanotomy, particularly in cases involving anomalies in the positioning of the sigmoid sinus and facial nerve. However, with advancements in visualization methods of bony and membranous inner ear structures, such procedures are now possible in select cases. Alternative surgical approaches to the inner ear have also been developed for cochlear implantation.

The issue of rehabilitating children and adolescents with hearing loss is increasingly important today due to the growing number of individuals with hearing impairments and the overall rise in congenital teratogenic conditions, particularly anomalies of the auditory system.

Currently, there are three levels of prevention for congenital and hereditary pathologies worldwide: primary, secondary, and tertiary. While the first two are typically the responsibility of pediatricians, geneticists, and other specialists, the tertiary level involves postnatal rehabilitation and corrective treatment, in which otologic surgeons and plastic surgeons play a key role.

Over the last decade, significant experience has been accumulated in reconstructive surgery for children with external and middle ear malformations. This remains a challenging and relevant issue in reconstructive otologic surgery.

Rehabilitating children and adolescents with hearing, speech, and intellectual developmental delays is of particular importance in the context of societal humanization efforts. It is known that hearing impairment leads to speech development delays, pronunciation issues, negatively affects cognitive development, and can cause persistent depression, leading to psychological disorders and developmental delays in children and adolescents. Therefore, the search for modern methods of reconstructive and rehabilitative surgery for the pathological auditory organ is of high relevance.

Given the growing incidence of children with congenital ear anomalies, the integration of audioprosthetics with surgical treatment involving geneticists, surdopedagogues, phoniatrists, psychologists, and neurologists is of great socio-psychological importance.

The first two years of life are critical for speech acquisition, with hearing playing a fundamental role in the development of verbal communication. The lack of auditory capacity can delay cognitive development, increase psychological issues, raise the risk of traumatic experiences, and contribute to illiteracy. Early medical intervention is therefore essential for children with hearing impairment. Children with microtia and atresia often suffer from low self-esteem and social dysfunction, requiring hearing rehabilitation and cosmetic reconstructive auricular surgery to improve appearance [15,16,17].

To date, bone conduction hearing aids remain one of the most reliable methods of auditory rehabilitation. These devices are attached to the head via a softband, transmitting sound signals directly to the inner ear through the skull bone. Implanted bone conduction hearing devices use titanium implants to connect a vibratory transducer to the skull via a subcutaneous abutment. The timing of the operation and the placement of the implant are critically important.

Therefore, auditory prosthetics in children with congenital anomalies of the outer and middle ear remain a pressing issue in modern otorhinolaryngology.

Study Objective: To evaluate the development and effectiveness of hearing in children with bilateral congenital aural atresia after using the Ponto Softband, and to compare their results with those of children with normal hearing.

Study Material: Our team studied the improvement in hearing in patients with bilateral microtia and atresia using Softband hearing aids. We recommend the early application of bone conduction hearing devices in newborns with bilateral microtia and atresia. There is currently no known developmental curve for hearing in such children using bone conduction hearing aids. Our aim was to evaluate auditory development in children with bilateral microtia and atresia using the Softband. Over a two-year period, we observed 40 children.

Main Group: 40 infants aged from 3 months to 2 years using a Ponto bone conduction hearing implant attached with a Softband. To assess the level of auditory integration, we used the "Infant-Toddler Meaningful Auditory Integration Scale" (IT-MAIS), evaluating hearing development at birth and again at 3, 6, 12, and 24 months. Visual reinforcement audiometry was used to assess auditory thresholds. High-resolution CT scans were used to evaluate the degree of hearing loss. Hearing thresholds and speech intelligibility indicators

were also measured.

All parents gave written consent for the use of the Softband and for participation in the study.

The Softband group included 40 infants with bilateral microtia and atresia (23 boys, 17 girls), aged between 3 months and 2 years (mean age 8.45±5.67 months; median 7 months). All patients used the Ponto device (Ponto Pro, produced by Oticon) with a Softband. Based on Marx's classification, all 40 patients were categorized as group III based on the degree of deformity and bilateral atresia. The average bone conduction threshold was 18.13±4.84 dB nHL (range 0–25 dB nHL), and the air conduction threshold was 76.38±4.53 dB nHL (range 70–85 dB nHL), as determined by auditory brainstem response [18,19,20].

Auditory and speech development in infants and toddlers was evaluated using the Chinese version of the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), which is a structured questionnaire for parents with 10 items covering three domains: vocal behavior (items 1–2), response to sound (items 3–6), and deriving meaning from sound (items 7–10). Each item had five response options: 0 = never, 1 = rarely, 2 = occasionally, 3 = frequently, and 4 = always. The IT-MAIS was administered at baseline (0 months) and at 6, 12, and 24 months after fitting the Ponto Softband.

Visual reinforcement audiometry was conducted during the first 6 months post-fitting to determine hearing thresholds. Tone audiometry was performed at 250, 500, 1000, 2000, 4000, and 8000 Hz (both pre- and post-surgical intervention). Speech perception tests were conducted in Chinese using recorded speech materials read twice aloud, with the average result used in final analysis.

All data were analyzed using Statistica 10.0. Continuous variables were expressed as mean ± standard deviation. IT-MAIS scores were compared to standard values. Repeated measures ANOVA was used to assess differences between standard values and IT-MAIS scores in Softband users. Paired t-tests were used to assess differences between groups with and without Softband. ANOVA was used to compare pre- and post-surgical outcomes in implant recipients. Values of $p < 0.01$ were considered statistically significant.

3. Discussion

The overall auditory integration according to the "Infant-Toddler Meaningful Auditory Integration Scale" (IT-MAIS) was assessed in infants and toddlers from three groups: those without surgery, those who showed significant improvement in hearing, and those with hearing thresholds within normal limits. At the initial stage, the average value of auditory thresholds using visual reinforcement audiometry with Softband hearing devices was 76.75±6.05 dB HL, and improved to 32.25±6.20 dB HL (P<0.01).

In the implantation group, the average hearing threshold at baseline, before and after surgical intervention, was 59.17±3.76 dB HL, 32.5±2.74 dB HL, and 17.5±5.24 dB HL, respectively (P<0.01). Speech recognition scores improved from 23.33±14.72% to 77.17±6.46%, and then to 96.50±2.66% (P<0.01).

In the Softband group of 40 children, auditory integration levels were assessed at baseline (0 months), and at 3, 6, 12, and 24 months. The mean follow-up period was 21.9±4.6 months. 17.5% of patients did not attend follow-up. On average, children wore the Softband device for 6.57±1.22 hours per day.

The overall IT-MAIS scores in the Softband group showed a significant improvement over time, though they remained below standard levels. The mean differences between the standard and Softband group scores at baseline and at 3, 6, 12, and 24 months were 44.30±15.31%, 38.15±12.87%, 29.18± 10.95%, 19.80±10.55%, and 5.24±11.55%, respectively. This difference was significant and decreased gradually over time (Greenhouse-Geisser method, $F = 144.09$, $P<0.001$).

At baseline, the mean aided hearing threshold measured by audiometry was 76.75 ± 6.05 dB HL; with the Ponto Softband device, the threshold improved to 32.26 ± 6.20 dB HL. A paired t-test showed a significant improvement of 44.50 ± 3.89 dB HL ($t = 72.36$, $P<0.01$).

The average speech recognition scores were 77.17 ± 6.46% in the Softband group and 96.50 ± 2.66% in the implanted Ponto group. These improvements, according to statistical data, corresponded to 53.84 ± 15.29% and 73.17 ± 15.29% ($F = 97.45$, $P < 0.001$). The mean unaided bone conduction threshold was 17.5 ± 5.9 dB nHL, air conduction threshold was 72.5 ± 9.3 dB nHL, and VRA threshold in five patients was 30.5 ± 5.9 dB nHL. The total IT-MAIS detection and discrimination scores especially improved after using the Ponto Softband to near-normal levels.

Auditory perception plays a critical role in early speech development. Limitation of verbal interaction may slow the development of neuro-linguistic brain regions responsible for grammar acquisition and foreign language learning. Bilateral microtia-atresia significantly impairs hearing and speech, creating barriers to communication. Recent studies confirm that children with microtia-atresia belong to a high-risk group with possible delays in speech, intellectual development, and behavioral issues affecting academic performance. Moreover, limited auditory stimulation (as indicated by a significant air-bone gap) may alter the development of the central auditory system.

There is broad consensus that early auditory rehabilitation supports auditory perception and speech development. To date, no long-term studies with large samples have evaluated speech development in children with bilateral microtia-atresia using Softband bone conduction hearing devices. In our study, we conducted a two-year follow-up of auditory perception development in 40 Chinese-speaking children with bilateral microtia-atresia using Softband-mounted bone conduction hearing devices. We found that the Softband significantly improved auditory perception development.

Parental questionnaires were the most effective tool to assess early prelingual development and intervention outcomes since children at this age are unable to perform standard auditory tests. We used the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), which is a validated tool for

evaluating auditory development and rehabilitation efficacy. For comparison, 120 infants from Chinese-speaking families with normal hearing were also evaluated for speech sound detection, recognition, and discrimination. According to the IT-MAIS, children with bilateral microtia-atresia demonstrated an average delay of 44.30±15.31% in auditory perception development, highlighting the need for early intervention.

Current treatment options for bilateral microtia-atresia are limited and include surgical reconstruction of the external auditory canal and implantation of bone conduction hearing devices. Both approaches improve speech perception. However, reconstructive surgery may result in complications such as facial nerve damage and canal stenosis. In patients with microtia-atresia, the second stage of surgery may lead to complications due to limited blood supply to the implanted flap.

Given the growing demand for auditory improvement and the risks associated with reconstructive surgery, we recommend the use of bone conduction hearing devices with Softband before auricular reconstruction in children with bilateral microtia-atresia.

4. Conclusions

The findings of this study confirm that early auditory rehabilitation using the Ponto Softband bone conduction hearing system is a safe, non-invasive, and effective method for improving auditory perception and speech development in infants and toddlers with bilateral microtia and congenital aural atresia. Significant improvements in hearing thresholds and IT-MAIS scores over the 24-month follow-up period demonstrate the device's ability to facilitate meaningful auditory integration and communication skills during critical stages of neurodevelopment.

The use of bone conduction devices prior to auricular reconstruction offers a reliable interim solution that avoids the risks associated with early surgical intervention. By enabling timely auditory stimulation, these devices contribute not only to language acquisition and cognitive function, but also to psychosocial well-being, self-esteem, and academic readiness in affected children.

Given the high incidence of developmental delays associated with untreated bilateral atresia, and the limited effectiveness of surgical options in infancy, this study supports the early and widespread adoption of Softband-mounted bone conduction systems as part of a comprehensive, multidisciplinary approach to pediatric hearing loss rehabilitation.

REFERENCES

[1] Andreas R. et al. Literature Review: Rehabilitation of Ear Congenital Abnormalities // European Journal of Theoretical and Applied Sciences. – 2023. – T. 1. – №. 3. – C. 191-195.

[2] Batuk M., Gündüz B., Fetoni A. R. Audiological Approach in Bone-Anchored Hearing Aid Applications // Neurotology Updates. – Cham: Springer Nature Switzerland, 2024. – C. 437-460.

[3] Bergman J. E. H. et al. Updated EUROCAT guidelines for classification of cases with congenital anomalies // Birth Defects Research. – 2024. – T. 116. – №. 2. – C. e2314.

[4] Cywka K. B., Król B., Skarżyński P. H. Effectiveness of bone conduction hearing aids in young children with congenital aural atresia and microtia // Medical Science Monitor: International Medical Journal of Experimental and Clinical Research. – 2021. – T. 27. – C. e933915-1.

[5] Frenzel H. Hearing Rehabilitation in Congenital Middle Ear // Advances in Hearing Rehabilitation. – 2018. – T. 81. – C. 32-42.

[6] Ito T. et al. Transcanal endoscopic ear surgery for congenital middle ear anomalies //Otology & Neurotology. – 2019. – T. 40. – №. 10. – C. 1299-1305.

[7] Kelley P. E., Scholes M. A. Microtia and congenital aural atresia // Otolaryngologic Clinics of North America. – 2007. – T. 40. – №. 1. – C. 61-80.

[8] Kesser B. W., Chang K. W. 197 Evaluation and Management of Congenital Aural Atresia // Cummings Pediatric Otolaryngology E-Book. – 2021. – C. 259.

[9] Kesser B. W., Goddard J. C. Congenital Malformation of the External Auditory Canal and Middle Ear // Otologic Surgery E-Book. – 2021. – C. 43.

[10] Korver A. M. H. et al. Congenital hearing loss // Nature reviews Disease primers. – 2017. – T. 3. – №. 1. – C. 1-17.

[11] Lee M. Y. et al. Current treatments for congenital aural atresia // Journal of Audiology & Otology. – 2020. – T. 24. – №. 4. – C. 161.

[12] Liaw J., Patel V. A., Carr M. M. Congenital anomalies of the external ear // Operative Techniques in Otolaryngology-Head and Neck Surgery. – 2017. – T. 28. – №. 2. – C. 72-76.

[13] Morrison D., Kesser B. Congenital anomalies of the ear canal // Otolaryngologic Clinics of North America. – 2023. – T. 56. – №. 5. – C. 933-948.

[14] Patel A. A. et al. Incidence of audiologic or otolaryngologic evaluation in patients with external ear anomalies // American Journal of Otolaryngology. – 2024. – T. 45. – №. 3. – C. 104199.

[15] Rooijers W. et al. Hearing impairment and ear anomalies in craniofacial microsomia: a systematic review // International Journal of Oral and Maxillofacial Surgery. – 2022. – T. 51. – №. 10. – C. 1296-1304.

[16] Van Maldergem L., Van Camp G., Deltenre P. Hearing impairment // Swaiman's Pediatric Neurology. Elsevier. – 2024. – C. 43-51.

[17] Wang Q. S. et al. Pure tone audiometry is a new method for evaluating congenital malformation of the middle and outer ear (CMMOE) // Acta Oto-Laryngologica. – 2023. – T. 143. – №. sup1. – C. S30-S33.

[18] Worrall D. M. et al. Bone Anchored Hearing in Children with Aural Atresia: A comparison of outcomes with Transcutaneous Magnetic Surgical and non-surgical options // Madridge J Otorhinolaryngol. – 2018. – T. 3. – №. 1. – C. 64-69.

[19] Yellon R. F., Denoyelle F. 15 Evaluation and Management of Congenital Aural Atresia // Cummings Pediatric Otolaryngology. – 2014. – С. 196.

[20] Zhou X. et al. Epidemiology of congenital malformations of the external ear in Hunan Province, China, from 2016 to 2020 // Medicine. – 2024. – Т. 103. – №. 15. – С. e37691.