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Case Report

Navigating Hearing Restoration in Treacher Collins Syndrome: A Comprehensive Review and Case Series on Baha Applications

Yavuz Gündoğdu, Orhan Asya, Badel Tereci Oran, Celal Emre Çavlan and Ali Cemal Yumuşakhuylu*

Department of Otorhinolaryngology-Head and Neck Surgery, Marmara University, Istanbul

Abstract

Treacher Collins syndrome (TCS) is a rare genetic disorder characterized by craniofacial anomalies and bilateral conductive hearing loss. Early hearing rehabilitation is crucial for normal speech and language development. Bone-anchored hearing aids (BAHA) offer a reliable solution, with advantages such as simplicity, low complication rates, and improved quality of life compared to traditional hearing aids.

This study retrospectively reviews four TCS patients who underwent BAHA implantation. All patients exhibited characteristic craniofacial features and conductive hearing loss. Post-implantation, hearing thresholds improved significantly, and speech development reached levels comparable to peers. No intraoperative or postoperative complications were observed, and a six-week osseointegration period was ensured before external device attachment.

BAHA devices bypass middle ear deformities, enabling effective sound transmission through osseointegration. This study highlights the importance of early intervention and supports BAHA as a safe and effective method for auditory rehabilitation in TCS patients.

Keywords: Treacher Collins Syndrome; Bone-Anchored Hearing Aid (BAHA); Conductive Hearing Loss (CHL); Auditory Rehabilitation, Craniofacial Anomalie

Introduction

Treacher Collins syndrome, also known as mandibulofacial dysostosis, is a genetic disorder characterized by hypoplasia of certain facial bones. Common manifestations of the syndrome include cleft palate, micrognathia, hypopharyngeal obstruction, microcephaly, limb anomalies, renal and cardiac abnormalities, intellectual disabilities, downward-slanting palpebral fissures, and lower eyelid colobomas [1].

Ear anomalies in TCS are typically bilateral. These patients may exhibit abnormal pinna positioning, microtia, atresia of the external auditory canal, middle ear deformities, and ossicular anomalies. Early rehabilitation is essential for patients with bilateral conductive hearing loss to support language development. Due to external auditory canal anomalies, these patients benefit more from bone-conduction hearing devices than from traditional hearing aids.

BAHA are an effective hearing rehabilitation method for children with Treacher Collins syndrome, with a relatively straightforward surgical procedure and low complication rates [2,3].

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***Corresponding author:** Gündoğdu Y, Department of Otorhinolaryngology-Head and Neck Surgery, Marmara University, Istanbul

Case Presentation

In this study, patients with TCS who underwent BAHA implantation at our department of otorhinolaryngology and head and neck surgery, were retrospectively reviewed (Table 1).

Case 1

HAK is a 6-year-old patient with congenital hearing loss. Diagnosed with TCS, the physical examination revealed bilateral atresia of the external auditory canals, drooping eyelids, micrognathia, zygomatic hypoplasia, and missing teeth. The patient had no additional medical conditions or history of previous surgeries. Audiometry results showed moderate bilateral CHL. Audiology follow-up indicated that the patient's speech and language development was comparable to peers.

At age 6, BAHA was implanted in the right ear at our clinic. No early or late postoperative complications were observed. The patient consistently uses the external BAHA component, except during sleep. A six-week period was allowed for osseointegration. Post-implantation audiometry revealed a hearing threshold of 18.75 and a speech recognition threshold of 20 dB. The patient reported that the device enhanced quality of life and was easy to use.

Preoperative and postoperative hearing thresholds are illustrated in Figure 1.

Case 2

SFY is a 20-year-old patient with congenital hearing loss and a diagnosis of TC. Physical examination revealed bilateral atresia of the external auditory canals, micrognathia, zygomatic hypoplasia, and pectus excavatum. Prior to using a bone-conduction device, the patient's family noticed developmental delays in speech and language compared to peers. Following device use, the patient's speech and language skills improved to a level similar to that of peers.

Table 1: Patient Characteristics.

Patient	Gender	Family History	Comorbidities	Previous Surgery	Age at Surgery	Side	Age of Softband Initiation	Neonatal Hearing Test
HAK	M	No	No	None	6	R	5.5 months	Bilateral Moderate CHL
SFY	M	No	No	Septorhinoplasty	20	R	5 years	*
EK	M	No	No	Cleft Palate repairment	10	L	6 months	Bilateral Moderate CHL
MSB	M	No	No	None	11	R	2 months	Bilateral Moderate CHL

* The patient's neonatal audiology data was unavailable.

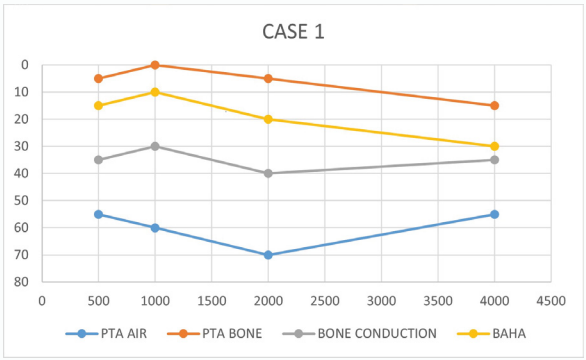

Figure 1: Preoperative and Postoperative Hearing Thresholds for Case 1.

At age 20, BAHA was implanted in the right ear at our clinic. Audiological data for the patient was unavailable. The patient reported experiencing intermittent redness and pain at the surgical site over a 2-3 year postoperative period, though no current complaints were noted. The patient wears the external BAHA component consistently, except during sleep. A six-week period was allowed for osseointegration.

Case 3

EK is a 10-year-old patient with congenital hearing loss and a diagnosis of TCS. Physical examination revealed bilateral atresia of the external auditory canals, drooping eyelids, micrognathia, and zygomatic hypoplasia. The patient also had a history of cleft palate repair. Audiometry results showed moderate bilateral CHL. Audiology follow-up indicated that the patient's speech and language development was at a level comparable to peers.

At age 10, BAHA was implanted in the left ear at our clinic. No postoperative complications were observed. The patient consistently wears the external BAHA component except during sleep. A six-week period was allowed for osseointegration. Post-implantation audiometry revealed a hearing threshold of 21.25 and a speech recognition threshold of 25 dB. The patient reported improved quality of life and ease of use with the device.

Preoperative and postoperative hearing thresholds are illustrated in Figure 2.

Case 4

MSB is an 11-year-old patient with congenital hearing loss and a diagnosis of TCS. Physical examination revealed bilateral atresia of the external auditory canals, drooping eyelids, micrognathia, and zygomatic hypoplasia. The patient began using a bone-conduction hearing device at 2 months of age. Audiology follow-up indicated that the patient's speech and language development was on par with peers.

At age 11, BAHA was implanted in the left ear at our clinic. No postoperative complications were observed. The patient consistently wears the external BAHA component except during sleep. A six-week period was allowed for osseointegration. Post-implantation audiometry showed a hearing threshold of 16.25 and a speech recognition threshold of 25 dB. The patient reported improved quality of life and ease of use with the device.

Preoperative and postoperative hearing thresholds are illustrated in Figure 3.

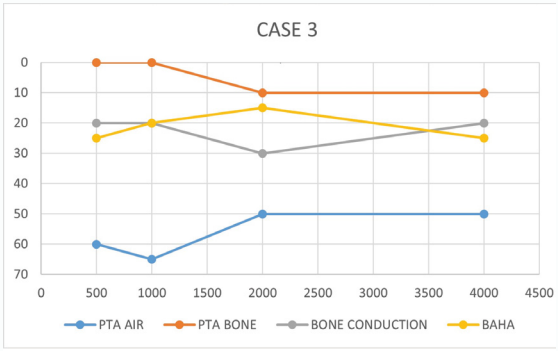

Figure 2: Preoperative and Postoperative Hearing Thresholds for Case 3.

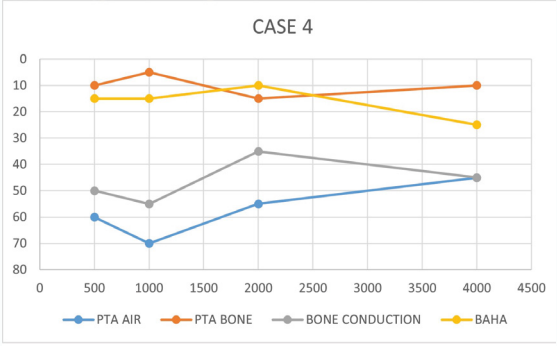

Figure 3: Preoperative and Postoperative Hearing Thresholds for Case 4.

Discussion

TCS is a syndrome that affects approximately one in 50,000 live births and is inherited in an autosomal dominant manner. Due to various ear anomalies, patients typically experience bilateral conductive hearing loss. For conductive hearing loss, traditional hearing aids, bone-conduction hearing devices, and bone-anchored hearing devices are options. In these patients, acromioplasty is one potential approach for hearing rehabilitation [4]. However, due to the risk of complications such as facial nerve injury and meatal stenosis, as well as the frequent need for revision surgeries, it is not commonly preferred [4,5]. Traditional hearing aids are generally not favoured in children with TCS due to external auditory canal anomalies.

To provide early hearing rehabilitation, the use of a soft band device is recommended in these patients at an early stage. These

devices can be used as early as three months of age, before the cranial bones have sufficiently thickened for implant surgery. After using the soft band bone-anchored hearing device for six months, most patients experience improvements in hearing rehabilitation, and their developmental progress approaches that of healthy infants and children. Despite these positive outcomes, some patients may experience aesthetic concerns, pressure-related headaches, or sores associated with the device [5]. Consequently, bone-anchored hearing implants are often preferred.

Advantages of BAHA include the simplicity of the surgical procedure, low complication rates, and applicability in the preschool years. TCS patients should start using BAHA at the earliest feasible age. However, early implantation is not recommended due to insufficient and irregular cranial bone thickness.

BAHA implantation is advised once the child's age and temporal bone thickness (≥ 3 mm) become suitable. In the United States and Canada, age (≥ 5 years) is considered, while in France, cortical thickness (≥ 3 mm) is taken into account. In our country, a minimum age of 5 years is also required for BAHA implantation.

BAHA was first developed in 1977. It consists of internal and external components: a titanium osseointegration implant is surgically placed on the cortex of the temporoparietal bone, located above and behind the external ear, and is connected to the external sound processor either directly via a percutaneous abutment or magnetically in transcutaneous systems. Sounds entering the external sound processor are converted into vibrations through the implant and surrounding bone, subsequently transmitting them to the cochlea in the inner ear.

Temporal bone CT should always be performed preoperatively to assess skull thickness and the positions of the facial and sigmoid sinuses.

Possible intraoperative complications include exposure of the dura mater and the sigmoid sinus. Potential postoperative complications involve flap necrosis or erythema, flap irritation or granulation, osseointegration failure, trauma, and surgical site infection. The implant loss rate is approximately 5%, with a slightly higher rate in children, ranging from 7.5% to 15%. Major complications, such as brain abscess, are rare [3].

The osseointegration process creates a direct structural and functional connection between the bone and implant surface without any intervening fibrous tissue. Osseointegration reflects the healing of the endosteum, involving stages such as hematoma formation, hematoma resolution, migration of osteogenic cells, and bone formation. The bone healing process around the implant mirrors physiological healing. After surgery, ankylosis forms at the contact point between the implant and bone surface, and this is maintained by a dynamic balance following osseointegration [3].

During surgery, the implant site is determined at the intersection of a horizontal line passing through the temporal line and an oblique line approximately 3,5 cm from the entrance of the external auditory canal. The thickness of the skin and subcutaneous tissue is measured. The incision area is marked, and the skin and subcutaneous layers are elevated supraperiosteally with an anterior base.

The implant site is prepared with a drill at the midpoint of the marked area, drilling down to the dura. A titanium screw is then implanted, and the flap is laid over the implant. Patients are discharged within 1-2 days post-surgery. The external component is attached six weeks later following osseointegration. In this study, no intraoperative or postoperative complications were observed in any of the patients.

In this retrospective study, audiology data and auditory rehabilitation in patients achieved the optimal level targeted post-implantation. BAHA implantation is a preferred surgical method due to its low complication rates and relatively simple surgical procedure.

The preoperative and postoperative aided thresholds of the patients are presented in Figure 4.

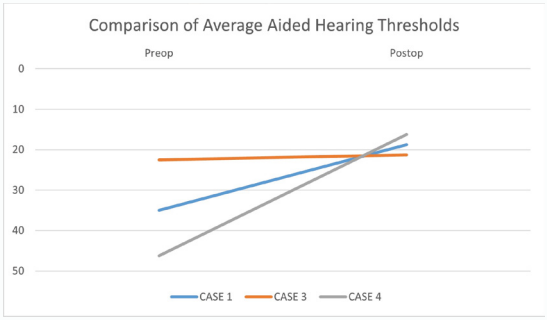

Figure 4: Comparison of Average Aided Hearing Thresholds for Our Patients.

A review of the literature indicates that bone conduction hearing devices are considered the most up-to-date method for treating conductive hearing loss in patients with TCS [1]. Early application of BAHA Soft band in these patients has been supported by numerous studies as an effective approach to achieving communication development goals [2-4]. The findings of our study are consistent with the existing literature, demonstrating that BAHA implantation represents an optimal option for TCS patients.

This study highlights the importance of early auditory rehabilitation in patients with TCS to support normal speech and language development. BAHA have proven to be a safe, effective, and reliable solution for managing conductive hearing loss in TCS patients. The surgical procedure is straightforward, with minimal risk of complications, and the functional outcomes significantly enhance the quality of life.

Our findings, consistent with the literature, underscore the critical role of BAHA in bypassing middle ear deformities and ensuring effective sound transmission via osseointegration. Early intervention with Soft band devices and timely BAHA implantation allows TCS patients to achieve developmental milestones comparable to their peers. Further studies with larger patient groups and long-term follow-ups are recommended to optimize surgical techniques and expand the clinical application of BAHA in craniofacial syndromes.

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