A Bone Conduction Implantable Device as a Functional Treatment Option in Unilateral Microtia with Bilateral Stapes Ankylosis: A Report of Two Cases

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Case series

Patient: Female, 29 • Male, 35

Final Diagnosis: Microtia with stapedial ankylosis

Symptoms: Hearing loss

Medication: —

Clinical Procedure: Bone conduction implantable device

Specialty: Audiology

Objective: Congenital defects/diseases

Background: Implantable devices have been proposed as an alternative to hearing aids and auditory canal reconstruction in patients with microtia (congenital aural atresia), which includes a malformation of the external and middle ear. This report is of two rare cases of microtia associated with congenital stapes ankylosis treated with an implantable device and describes the treatment outcomes.

Case Report: Two siblings from Ecuador, a 29-year-old woman, and her 35-year-old brother, were born with unilateral type II microtia with bilateral external auditory canal atresia and conductive hearing loss. Pre-operatively, high-resolution computed tomography (HRCT) imaging was performed using FastView software to allow placement of a bone conduction-floating mass transducer (BC-FMT) to couple a Bonebridge bone conduction implant (BCI) system in both patients. Pure-tone audiometry (PTA) testing and speech audiometry were performed. The Abbreviated Profile of Hearing Aid Benefit (APHAB) and the Speech, Spatial and Qualities (SSQ) of hearing scale questionnaires and scoring systems were used. Following activation of the implantable device, both patients achieved improved bilateral conductive hearing with sound-field (field-free) thresholds >25 dB, and speech recognition scores >90%. In both cases, hearing improvement remained at three years following surgery.

Conclusions: To our knowledge, these are the first reported cases of microtia with congenital stapes ankylosis successfully treated with a bone conduction implantable device. Patients with microtia and stapes ankylosis who are reluctant to undergo surgery may benefit from unilateral or bilateral, short-term or long-term use of a Bonebridge bone conduction implantable device.

MeSH Keywords: Bone Conduction • Congenital Abnormalities • Correction of Hearing Impairment • Ear, External • Hearing Aids • Hearing Loss, Bilateral

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Background

Congenital conductive hearing loss caused by malformations of the middle ear or external auditory canal can be treated with complex surgical procedures, or with bone conductive hearing aids that are either mounted on spectacles or implanted. Microtia, or congenital aural atresia, includes a malformation of the external and middle ear. The traditional surgical approach for microtia combines a wide canaloplasty and a tympanoplasty. However, these procedures involve a significant risk of damaging an aberrant facial nerve. Also, functional auditory gains with ossicular reconstruction are usually lower than in post-traumatic hearing loss or following chronic otitis, and hearing improvements are often short lasting due to fibrosis and re-stenosis of the external auditory canal. Conventional bone conductive hearing aids have the limitations of fluctuations of hearing performance if the contact is loose, or discomfort, inflammation, or ulceration of the skin at the contact site if it is too tight [1–4].

Bone conduction implantable hearing devices have been proposed as an alternative to hearing aids and auditory canal reconstruction in patients with microtia and provide optimal stability of the connection between the implant and bone with improved functional results when compared with conventional bone conductive hearing aids [3,5–8]. Bone conduction implantable hearing devices are classified as percutaneous or transcutaneous.

Percutaneous bone conduction implantable hearing devices involve a pillar screwed into the bone and abutting across the skin surface. Although major complications are very rare, skin reactions and infection at the implant site have been observed in between 2.4–38.1% of cases; failure of osseointegration and extrusion of the implant has been reported in up to 18% of cases in adults and 14.3% of cases in children, with a total rate of revisions ranging from 1.6–17.4% in adults and from 0–25% in children [4].

Transcutaneous bone conduction implantable hearing devices preserve the integrity of the skin, because the external part, the audio processor, is connected to the sub-cutaneous implant through coupled magnets. Candidates for bone conduction hearing implantable devices should be carefully selected to avoid unsuccessful implantation, and the different available options should be thoroughly discussed with the patient [9].

The Bonebridge active bone conduction prosthesis, or bone conduction implant (BCI) system, (MED-EL, Innsbruck, Austria) is a transcutaneous bone conduction hearing implantable device that requires the subcutaneous placement of a receiver and stimulator, connected to a vibrating module, the bone conduction-floating mass transducer (BC-FMT), drilled into the mastoid bone and fixed to its edges by two screws. Since 2011, the Bonebridge BCI system has been implanted in patients with conductive or mixed hearing loss [6,10–17]. In congenital atresia, or microtia, the Bonebridge BCI system provides a safe surgical approach, in the presence of lateral location of the facial nerve, sclerotic mastoid air cells, and a narrow facial recess space, and has the advantage of preserving the intact skin. However, in some cases, the surgical procedure for implantation of the Bonebridge BCI cannot be done without exposing the dura mater or ethmoid sinus and, in some patients with congenital aural atresia, instability of the ossicular chain might be present resulting in a reduction in bone conduction [18].

This report presents the cases of two siblings born with a unilateral type II microtia and bilateral conductive hearing loss who underwent a Bonebridge BCI and discuss the long-term functional outcomes and surgical pitfalls. In these two cases, the congenital bilateral defects were characterized by the rare association of microtia with congenital stapes ankylosis.

Case Report

Two Hispanic siblings from Ecuador, a 29-year-old woman, and her 35-year-old brother were born with the same physical features of unilateral type II microtia with atresia of the external auditory canal and bilateral conductive hearing loss. Their mother was born with bilateral conductive hearing loss, and at 25 years-of-age, her hearing failed to improve after stapedotomy for suspected otosclerosis, and she had then suffered a transient facial paresis and prolonged vertigo.

The two siblings had reported a subjective benefit in hearing after two months of use of conventional bone conductive hearing aids mounted onto their spectacles, but experienced difficulty in wearing these due to their microtia. Furthermore, they had no visual loss and complained of having to wear glasses with neutral lenses. Both patients underwent physical examination with otomicroscopy that showed a unilateral abnormal shape of the auricle and absence of the external ear canal on the right side for the female sibling, and on the left side for the male sibling, the pinnae in both patients appeared normal on the contralateral side, with an open, although small size, external auditory canal and an eardrum that was regular in appearance.

The patients underwent pure-tone audiometry (PTA) with average estimations at 0.5, 1, 2, and 4 KHz, using a Clinical Audiometer r37a (Resonance, Gazzangia, Italy). Middle ear impedance testing in the normally shaped ear was performed using a Clarinet Middle Ear Analyzer (Inventis, Padova, Italy) and showed a type A tympanogram with the absence of ipsilateral
stapedial reflexes. Speech perception tests included air-conducted, and bone-conducted word recognition scores (WRS) obtained in a free-field in a quiet sound booth. The audiological assessment was repeated on days 1, 3, 6, 12, and at 24 months, and 36 months following the activation of the Bonebridge BCI system. The Abbreviated Profile of Hearing Aid Benefit (APHAB) [19], and the Speech, Spatial, and Qualities (SSQ) hearing scale questionnaires [20], were administered pre-operatively, and at 6, 24 and 36 months, post-activation of the Bonebridge BCI system. The clinical and functional features for both patients are summarized in Table 1.

A high-resolution computed tomography (HRCT) scan of the temporal bones was then performed in both cases. A simulation of the application of the Bonebridge BCI system was performed in three-dimensions (axial, coronal, sagittal) with the Bonebridge BCI Fast View software (MED-EL, Innsbruck). The

| Patient 1. 29-year-old woman | Patient 2. 35-year-old man |
|-------------------------------|----------------------------|
| **Clinical features**         |                            |
| Right microtia and EAC atresia| Left microtia and EAC atresia|
| Previous procedures           | Unsuccessful left stapedoplasty for congenital malformation |
| Active Middle Ear Implant Score | 10 out of 16 |
| Duration of surgery/ complications | 38 minutes/none |
| PTA/ABG (dB HL)               |                            |
| Right: PTA=60, ABG=42.5       | Left: PTA=62.5, ABG=37     |
| Post-op.                       |                            |
| Right: PTA=19.5, ABG=2.0      | Left: PTA=24.5, ABG=5.5    |
| Contralateral PTA/ABG (dB HL) |                            |
| Left: PTA=52.5, ABG=33.5     | Right: PTA=55, ABG=33      |
| Pre-op. speech perception (WRS) |                            |
| Right: AC=100% at 85 dB HL, BC=100% at 20 dB HL | Right: AC=100% at 90 dB HL, BC=100% at 20 dB HL |
| Left: AC=100% at 70 dB HL, BC=100% at 25 dB HL | Left: AC=100% at 65 dB HL, BC=100% at 25 dB HL |
| Max. WRS in noise (free field): AC=55%, BC=75% | Max. WRS in noise (free field): AC=60%, BC=78% |
| Post-op. (3 months) speech perception (WRS in free-field) |                            |
| AC=100% at 25 dB HL           | AC=100% at 20 dB HL        |
| In noise: AC=80% at 45 dB HL   | In noise: AC=WRS 85% at 55 dB HL |
| Post-op. (36 months) speech perception (WRS in free-field) |                            |
| AC=100% at 25 dB HL           | AC=100% at 20 dB HL        |
| In noise: AC=88% at 40 dB HL   | In noise: AC=WRS 85% at 50 dB HL |
| APHAB                          |                            |
| Pre-op.                        | Post-op.                   |
| EC 16.3                        | 8.3                        |
| BN 33.3                        | 12.0                       |
| AV 18.5                        | 1.0                        |
| RV 31.3                        | 30.8                       |
| Pre-op.                        | Post-op.                   |
| EC 25.0                        | 8.3                        |
| BN 25.0                        | 12.0                       |
| AV 30.0                        | 11.0                       |
| RV 31.0                        | 30.2                       |
| SSQ scores                    |                            |
| Speech 1.4±1.8                 | 7.6±2.0                    |
| Spatial 0.5±2.0                | 3.2±4.0                    |
| Qualities 2.6±3.3              | 6.8±3.2                    |
| Pre-op.                        | Post-op.                   |
| Speech 2.0 ± 1.0               | 7.3 ± 1.9                  |
| Spatial 2.4 ± 0.8              | 6.5 ± 2.0                  |
| Qualities 1.8 ± 1.0            | 7.5 ± 2.0                  |

PTA – pure-tone average (0.5, 1, 2, or 4 kHz); ABG – air-bone gap; SDS – speech discrimination score; AC=WRS – air-conducted words recognition score; BC=WRS – bone-conducted words recognition score; pre-op – pre-operative (one-month before surgery); post-op – post-operative (at three and 36 months); SSQ – Speech, Spatial and Qualities (questionnaire); APHAB – Abbreviated Profile of Hearing Aid Benefit; EC – ease of communication (speech understanding under relatively favorable conditions); RV – reverberation (communication in reverberant settings); BN – background noise (communication in noisy settings); AV , aversiveness (unpleasantness of environmental sounds).
The distances between the bone conduction-floating mass transducer (BC-FMT) and the ethmoid sinus, the dura mater of the middle cranial fossa, the bony wall of the external auditory canal, and the Eustachian canal were calculated. (Figure 1) According to the Active Middle Ear Implant (AMEI) score, devised by Frenzel et al. [21], both patients were good candidates to an AMEI or a mastoid bone implant, with only moderate surgical risk. However, both patients preferred not to undergo a
staged mastoidectomy with tympanoplasty and stapedial surgery in order to avoid the risk of facial nerve damage.

Both patients underwent a unilateral Bonebridge BCI. The correct positioning of the BC-FMT was assessed intraoperatively by superimposing the captured correctly sized images on the CT images in the sagittal, frontal and axial planes using the dedicated Bonebridge BCI device system FastView software, a procedure similar to that described by Plontke et al. (Figure 2) [22]. In both patients, a pre-ethmoid location was chosen. As expected from the pre-operative analysis of the CT scan images, in both patients there was no need to expose the dura of the middle cranial fossa dura, nor the bony shell of the ethmoid sinus. Thinning of the bony wall of the external auditory canal was the only required additional step to gain sufficient room to fit the BC-FMT.

On the first post-operative day, a bone conduction threshold was obtained to ascertain that no sensorineural deterioration had occurred after the operation. The average bone conduction improved in both cases by up to 2.5 dB in the conductive hearing and up to 3.5 dB in conductive hearing, respectively. Direct stimulation of the implant was also performed to check the threshold of pure tones perceived by the patients by pure-tone audiometry (PTA) testing. Speech audiology was also performed.

At implant activation, one month later, an initial amplification response curve was created and stored in the audio processor using the Connexx Software. A subjective patient hearing threshold via the audio processor, combined with the pre-operative bone conduction threshold, allowed an implant functional threshold to be obtained called the Bonebridge BCI device Vibrogram, which acted as a calibration system for the fitting procedure. Two additional programs were then shaped according to the patient preferences and lifestyles.

At follow-up, 36 months following implant activation, the PTA, and the speech perception results were stable (Table 1). For both patients, the subjective benefit, measured by the APHAB questionnaire was particularly evident for the categories of ease of communication, listening in with background noise, and aversion to sound subscales; the subjective benefit, measured by the APHAB questionnaire, was not significant for the reverberation subscale. The SSQ hearing scale questionnaires showed a significant improvement in the pre-implant and post-implant hearing in all three domains, being particularly evident in improved speech discrimination in noisy situations (SSQ item numbers: 7 and 11) for both patients. The female patient also reported a reduction in effort to switch attention from one talker to another and in other similar tasks that implied selective attention (SSQ item numbers: 8, 9, 10, 12 and 14). The improvements in spatial localization were less prominent than the increase in speech perception in noise and the quality of the perceived sound.

Discussion

Microtia is often associated with atresia of the external auditory canal and with congenital anomalies of the middle ear [23]. Microtia is more frequently bilateral [24], leading to conductive hearing loss. The pinna in microtia is unable to support bone conductive hearing aids without discomfort.

In both patients presented in this report, high-resolution computed tomography (HRCT) imaging of the temporal bone

Figure 2. Intraoperative image of the Bonebridge bone conduction implant (BCI) system positioned in its mastoid well. Patient 2 (A). Note the two lateral fixation screws and the posteriorly seated receiver/stimulator, lodged in the sub-periosteal pocket. The implant shape (red) is superimposed on the three-dimensional (3-D) high-resolution computed tomography (HRCT) scan image for comparison (B).
showed unilateral bony atresia, with an apparently normal middle ear cavity, except for a dehiscent Eustachian canal in the tympanic segment. According to the established classification system, which addressed the feasibility of hearing restoration with conventional tympanoplasty and canaloplasty, both patients were poor candidates for this form of treatment [25]. When the more recently developed Active Middle Ear Implant (AMEI) score was applied to the findings in the two patients, which also took into account the accessibility of the oval and round window, the facial nerve displacement, and the pneumatized mastoid air cells, both patients were assessed as good candidates for implants. The unique nature of these two cases was the association of unilateral atresia of the external auditory canal and bilateral stapes ankylosis, and microtia occurred in the opposite ears in the two siblings. To our knowledge, these are the first reported cases of microtia with congenital stapes ankylosis.

In 2015, Kim described a case of bilateral oval window atresia but without atresia of the external auditory canal or microtia in an 8-year-old girl who received a Bonebridge bone conduction implant (BCI) system, which resulted significantly improved hearing [26]. This previous report confirms the long-term outcomes in cases with congenital ossicular anomalies, where the auditory ossicle system is not functioning, treated with Bonebridge BCI [26]. The safety and efficacy of the Bonebridge BCI system were initially addressed in a European multicenter study [2,28], and subsequently also demonstrated in children [29]. Riss et al. reported the outcome of the use of the Bonebridge BCI in 12 cases of atresia of the external auditory canal but recommended that implantation should be avoided if the bone conduction pure-tone threshold was less than 45 dB in conductive hearing loss [13].

In the two cases reported, the main reasons to select a Bonebridge BCI system were dependent on the anatomy of the temporal bones, which showed well pneumatized mastoid air cells, allowing for placement of the bone conduction-floating mass transducer (BC-FMT) at a safe distance (at least 2 mm) from all the relevant anatomical structures, including the middle cranial fossa, the dura, the ethmoid sinus, the Eustachian canal, and the bony wall of the external auditory canal. In accordance with the previously published literature, correct pre-operative assessment using three-dimensional (3D) reconstruction from high-resolution computed tomography (HRCT) is required for the optimal location for the BC-FMT [17]. However, there now exists a newly-designed surface template-assisted marker positioning device for the optimal location for the BC-FMT, but this was not used in these two cases [27].

The use of the bone conduction implant (BCI) system guarantees an optimal connection between the implant and the bone by osseo-integration [30,31], which usually occurs between four and 12 weeks postoperatively [32]. The percutaneous implant appears to offer better sound transmission compared with transcutaneous systems [28,33], but expose the recipient to infectious complications at the implant site [33]. The latter, instead, preserve the integrity of the skin and avoid complications; however, the sound transmission is partially dampened by the skin and subcutaneous layers, causing a loss, on average, of 5–7 dB conductive hearing loss. [35,36] The Bonebridge BCI system is a powerful enough to improve 55–71 dB hearing loss of amplification, at different frequencies [2,37–39]. In the two cases in this report, the pure-tone audiometry (PTA) was 60 dB and 62 dB, respectively, and the air-bone gap (ABG) were not within the normal range. Therefore, for these two cases, the Bonebridge BCI system was preferred to other forms of implantable hearing device.

A further area of interest of this case report was the immediate and persistent audiological benefit, despite both recipients being adults with a long-standing (congenital) conductive hearing loss. Also, no specific rehabilitation was needed, and soon after activation and fitting of the audio processor, both patients were able to understand a conversation, even in a non-native language. Importantly, the functional gain in hearing following implantation was comparable to previously published cases in the literature in terms of pure-tone thresholds (+39.25 vs. +26.1±13.7; 29.3±20.7) [40,41], whereas speech recognition scores were higher(100% at +70 dB, vs. +36.25 dB and 95% at +21%) [28]. The 2.5 dB hearing loss and 3.5 dB hearing loss bone conduction improvement observed in both cases was not surprising, given the known Carhart effect due to stapes ankylosis in congenital middle ear malformation [42].

The audiological results in the two cases observed from the scores of the Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaires, were similar to the recent findings of Monini et al. [43]. As far as we know, there have no previously published reports that have applied and compared the Speech, Spatial and Qualities (SSQ). However, in contrast with the findings of Weiss et al., [41], the two patients reported, showed an improvement in spatial localization with the Bonebridge BCI system, but this finding was less prominent than the increase in perception of speech and noise and the quality of the perceived sound. However, the use of subjective patient questionnaires for treatment response is a limitation in the assessment of the responses to the use of the Bonebridge BCI system in these two cases.

**Conclusions**

This report has described two cases of microtia with congenital stapes ankylosis, treated with a Bonebridge bone conduction implant (BCI) system, which provided long-term benefit...
for the hearing of both patients, without complications. The Bonebridge BCI device is suitable for patients well pneumatized mastoid air cells. Pre-operative planning should always include a three-dimensional (3-D) high-resolution computed tomography (HRCT) scan with superimposition of the shape of the bone conduction-floating mass transducer (BC-FMT), to select the most appropriate location for the BCI.

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Conflict of interest

None.
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