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Aesthetic and hearing rehabilitation in patients with bilateral microtia-atresia

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ABSTRACT

Objectives: To evaluate the safety and efficacy of auricle reconstruction and active transcutaneous bone-conduction implantation in patients with bilateral microtia-atresia.
Design: Patients were chosen prospectively, with each being his/her own control.
Setting: The setting was a tertiary referral center.
Participants: Twelve patients, aged 6–18 years, with bilateral microtia-atresia suffering from bilateral conductive hearing loss. All had an upper bone conduction threshold limit of 45 dB HL at frequencies of 0.5–4 kHz.
Main outcome measures: Patient satisfaction with the reconstructed auricle was rated as highly satisfactory, basically satisfactory, or unsatisfactory. Mean pure-tone thresholds and speech audiometry test results were compared among patients unaided, with a soft-band Bonebridge, and with an implanted Bonebridge. Subjective satisfaction was analyzed using three questionnaires: the Abbreviated Profile of Hearing Aid Benefit (APHAB), the Glasgow children's benefit inventory (GCBI), and the International Outcome Inventory for Hearing Aids (IOI-HA).
Results: All patients who underwent auricle reconstruction expressed satisfaction with their appearance. The mean pure-tone thresholds of unaided patients and those with soft-band and implanted Bonebridge were 55.25 ± 3.43 dBHL, 31.37 ± 3.03 dBHL, and 21.25 ± 2.16 dBHL, respectively. The mean speech discrimination scores measured in a sound field with a presentation level of 65 dB SPL under these three conditions were 46.0 ± 0.11%, 80.0 ± 0.09%, and 94.0 ± 0.02%, respectively. Questionnaires demonstrated patients' benefits and satisfaction with this surgery.
Conclusions: The surgical procedure involving auricle reconstruction and Bonebridge implantation was safe and effective for patients with bilateral microtia-atresia, solving both appearance and hearing problems.

1. Introduction

Microtia-atresia is characterized by abnormalities of the auricle (microtia) and aplasia or hypoplasia of the external auditory canal, often associated with middle ear abnormalities. The incidence of congenital microtia-atresia has been estimated to be one in 10,000 births, with about one-quarter being bilateral [1]. This condition is frequently associated with various syndromes, including Treacher-Collins, Goldenhar syndromes and hemifacial microsomia [2]. Congenital microtia-atresia affects patients in two specific ways. The first is severe conductive hearing loss (HL) with an air-bone gap of 50–60 dB, which, if not corrected in a timely manner, may delay speech development. The second is feelings of inferiority and problems integrating into social environments due to auricle malformation [3]. Resolution of microtia-atresia therefore requires both hearing rehabilitation and appearance improvement.

Patient appearance may be improved by two-stage auricle reconstruction surgery, including implantation of a skin soft tissue expander and auricle reconstruction using autogenous rib cartilage.

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These patients also require hearing rehabilitation to improve their hearing. Methods of hearing rehabilitation include surgical reconstruction of the external aural canal (atresiaplasty), conventional bone conduction hearing aids, and percutaneous bone anchored hearing aids (BAHA). Despite good rehabilitation results, these procedures have been associated with various complications, including canal restenosis, chronic infections, salivary fistula, local inflammation, skin-overgrowth, and implant extrusion [4,5]. Although transcutaneous passive skin-drive magnetic devices were developed to overcome the limitations of percutaneous bone anchored hearing aids, these devices have the disadvantage of a transcutaneous attenuation of 10–15 dB [6].

The Bonebridge is an active transcutaneous bone-conduction implant (MED-EL Corporation, Innsbruck, Austria) which consists of two major parts, a magnetic implant and an external audio processor. The external processor provides active direct-drive transcutaneous conduction to the magnetic receiver under the skin, directly stimulating the bone via an electromagnetic transducer screwed onto the mastoid [7]. This system was found to have fewer complications compared with percutaneous bone conduction implants and showed proven auditory benefits [8]. Therefore, unilateral Bonebridge should theoretically benefit patients with bilateral microtia-atresia.

Determining the position of the Bonebridge implant is important, as the audio processor should not interfere with auricle reconstruction. If the audio processor touches the skin flaps or ear rim, it can potentially impair the blood supply to the reconstructed auricle, which may lead to necrosis in the reconstructed ear and produce acoustic feedback [9]. A three-stage surgical procedure involving auricle reconstruction and Bonebridge implantation was therefore developed. The Bonebridge implantation was performed after the auricle was reconstructed, or before implantation of the skin soft tissue expander. The first aim of this study was to describe the surgical procedure of Bonebridge implantation.

Although Bonebridge implantation has shown good outcomes [3,7,8,10], few studies have assessed the efficacy of Bonebridge in speakers of Mandarin. The second aim of this study was to evaluate the benefits of unilateral Bonebridge implantation, using Mandarin Speech Test Materials (MSTMs) and three questionnaires, in 12 Mandarin-speaking patients with bilateral microtia-atresia treated at Peking Union Medical College Hospital (PUMCH).

2. Materials and methods

2.1. Participants

This single center prospective study involved patients with bilateral microtia-atresia treated at PUMCH in Beijing, China, and was approved by the Institutional Review Board of PUMCH. Patients were included if they were aged >6 years, of height >1.28 m, had bone conduction hearing thresholds >45 dB HL at frequencies of 0.5–4 kHz, and were psychologically and emotionally stable. Patients with unilateral microtia-atresia, malformation of the inner ear (sensorineural hearing loss) or concomitant diagnosed conditions such as cerebral palsy and intellectual disability were excluded from this study.

Twelve patients (Nine boys, three girls) with bilateral microtia-atresia were enrolled in the study. Degrees of auricular dysplasias were evaluated according to Max's classification [11]. Their mean (SD) age was 11.0 (5.0) years (range, 6–18 years). All had conductive hearing loss due to bilateral atresia. All had used a soft-band Bonebridge for at least 6 months prior to unilateral Bonebridge implantation surgery, performed at PUMCH between March 2016 and October 2016. The detailed characteristics of these patients are shown in Table 1.

2.2. Surgical techniques involving auricle reconstruction and Bonebridge implantation

Before Bonebridge implantation, patients underwent a temporal bone high-resolution computed tomography (HRCT) scan to evaluate the structure of their temporal bone and middle ear. These CT datasets were imported into the three-dimensional simulation software to determine the optimal site of the Bonebridge-FMT (Fig. 1) [12]. Patients were graded by the Jahrsdoerfer grading scale [13]. Based on the results of three-dimensional simulations, all patients underwent Bonebridge implantation via a transmastoid (TM) approach.

Patients who wish aesthetic and hearing rehabilitation were performed three-stage surgery. During the first stage, a soft skin expander was implanted in the mastoid region. During the second stage, the auricle was reconstructed using autogenous rib cartilage. During the third stage, performed 6 months later, the Bonebridge was implanted. The surgical procedures are shown in Fig. 2.

Table 1
Demographics and clinical data of patients.

Item	Gender†	Age (years)†	Degree of auricular-microtia (right ear/left ear)*‡	Syndrome	Performed surgery Before BB implantation	BB implantation pre or post auricle reconstruction	Js Grading
1	M	6.5	III/III		Left auricle reconstruction + right skin expander implantation	post	6
2	M	9	III/II		Right auricle reconstruction + left Vibrant Sound-bridge implantation	post	6
3	M	7.5	III/III		Right auricle reconstruction + right atresiaplasty	post	7
4	F	6	II/III	Goldenhar	none	pre	6
5	M	7	III/III		Right auricle reconstruction + left skin expander implantation	post	5
6	M	7	III/III		Bilateral auricle reconstruction	post	6
7	F	18	III/III		Right atresiaplasty	post	7
8	M	6	II/III		none	pre	6
9	M	18	III/II		Right skin expander implantation	post	6
10	F	13	III/III		none	pre	6
11	F	16	III/III		none	post	6
12	F	18	III/III		none	post	6

†F, female; M, male; Age, age in years at the time of Bonebridge implantation.
‡Degrees of auricular dysplasias were evaluated according to Max's classification.