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Case Report

Bonebridge Implantation for Conductive Hearing Loss in a Patient with Oval Window Atresia

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The occurrence of oval window atresia is a rare anomaly with conductive hearing loss. Traditional atresia surgeries involve challenging surgical techniques with risks of irreversible inner ear damage. Recent reports on Bonebridge (Medel, Innsbruck, Austria), a novel implantable bone conduction hearing aid system, assert that the device is safe and effective for conductive hearing loss. We present a case of Bonebridge implantation in an eight-year-old girl with bilateral oval window atresia.

KEYWORDS: Oval window atresia, conductive hearing loss, Bonebridge

INTRODUCTION

Congenital anomalies of the oval window in the ear are rare and have been reported in 0.5–1.2% of patients with pediatric conductive hearing loss [1]. Both hearing aids and surgical approaches have been used for treatment, including vestibulotomy [2], fenestration surgery of the lateral semicircular canal [3], and malleostapedotomy [4]. However, these surgical treatments are technically difficult, and the risks of inner ear damage and subsequent sensorineural hearing loss are not insignificant [5]. In some cases, an accompanying abnormal facial nerve course aborts surgical approaches [6].

Recently, a novel active bone conduction hearing aid—Bonebridge implant system (Medel, Innsbruck, Austria)—was developed for the treatment of conductive hearing loss. Favorable hearing outcomes have been reported with the use of Bonebridge in patients with conductive hearing loss [7, 8]. However, there have been no reports of Bonebridge implantation for patients with conductive hearing loss due to oval window atresia.

Here we present a case of Bonebridge implantation for the treatment of conductive hearing loss in a patient with bilateral oval window atresia.

CASE PRESENTATION

An eight-year-old girl was referred to our clinic due to bilateral congenital hearing loss. She had a history of using conventional hearing aids but complained of ear fullness, low speech discrimination, and poor sound quality. On admission to our clinic, the patient showed no symptoms of dizziness or facial palsy. Her initial pure tone audiometry demonstrated bilateral, moderate–severe, and conductive hearing loss with a mean air–bone gap of 37 dB in the right ear and 49 dB in the left ear (Figure 1). The patient's speech recognition threshold (SRT) was 60 dB in the right ear and 66 dB in the left ear, and her speech discrimination score (SDS) was 92% at the most comfortable level (MCL) of 90 dB, bilaterally.

Temporal bone computed tomography showed the absence of stapes and oval windows and also revealed an abnormal facial nerve course running inferior–medially over the oval window areas of the patient on both sides (Figure 2).

Under general anesthesia, retroauricular skin incision and periosteal flap elevation were performed on the left side. A bony well for the Bonebridge transducer was created in the mastoid area, and a posterior subperiosteal pocket for the demodulator and magnet was made. The internal device was firmly fixed with two screws. Postoperative transorbital X-ray confirmed the appropriate position of the device, with no penetration of the dura (Figure 3 a, b). No intraoperative or postoperative complications occurred.

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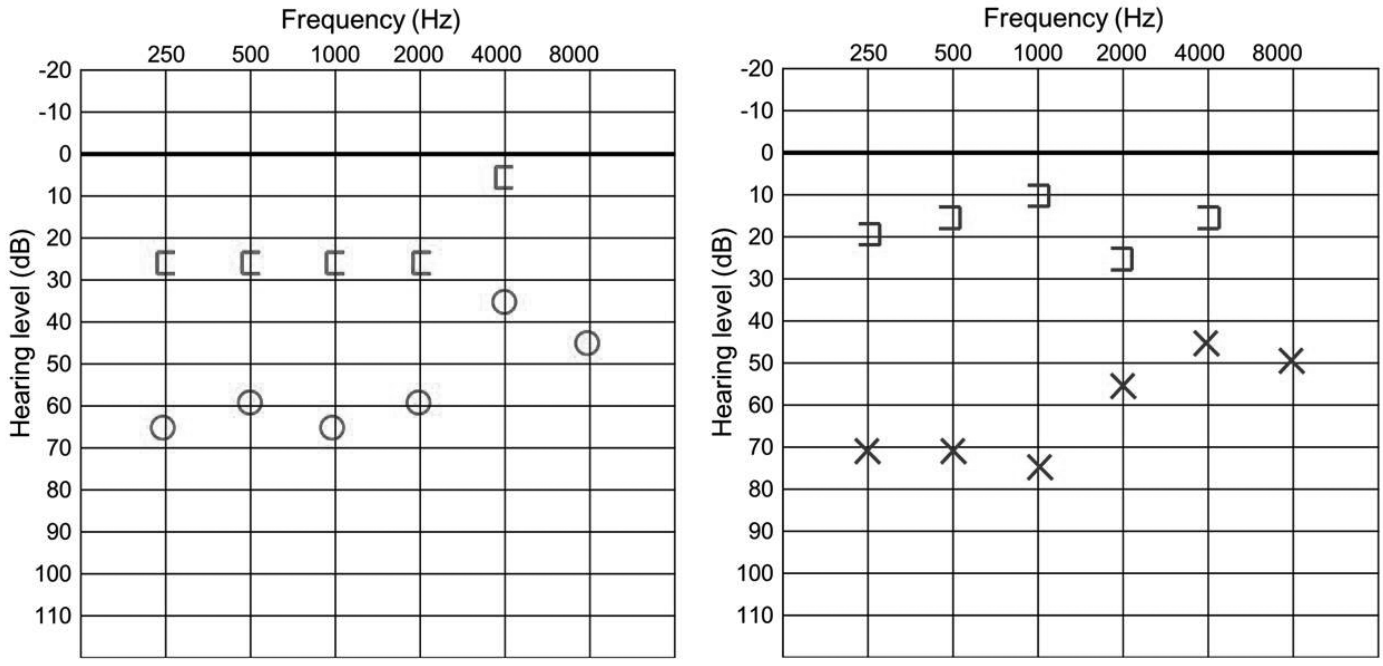

Figure 1. Initial pure tone audiogram

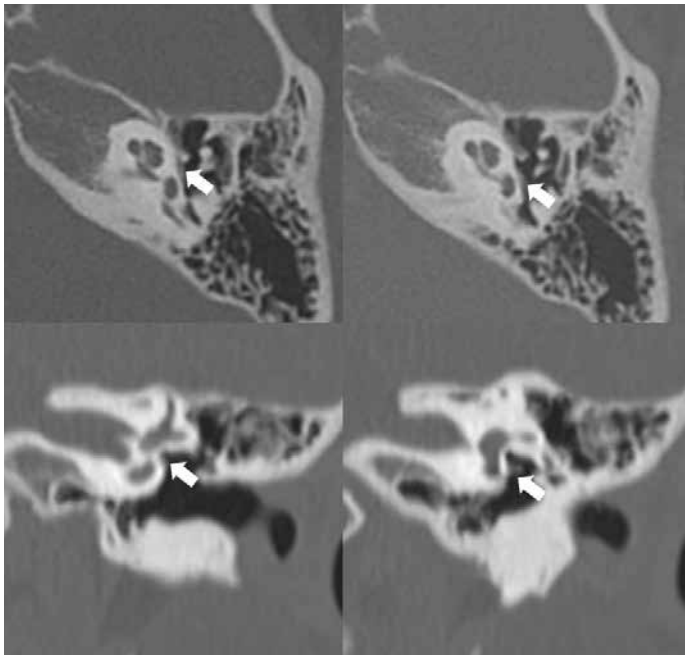

Figure 2. Temporal bone computed tomography. Inferior–medial displacement of the facial nerve (white arrow) and absence of both stapes and oval window

The device was activated after four weeks without any problems with the wound. The patient was satisfied with the clear sound quality immediately after the device was switched on. Three months after the insertion of the device, the pure tone threshold in the presence of contralateral noise was 33 dB in the right ear and 25 dB in the left ear in the free-field condition (Figure 4). Speech tests revealed SRT of 22 dB in the right ear and 18 dB in the left ear and SDS of 100% at MCL of 50 dB, bilaterally. No written informed consent was obtained as this was a case report preserving anonymity without recognizable photographs of the patient.

DISCUSSION

There are several surgical treatments for conductive hearing loss due to oval window atresia, including vestibulotomy, fenestration surgery of the lateral semicircular canal, and malleostapedotomy. These surgeries are not only technically challenging but also show unreliable postoperative hearing outcomes [2-4]. In particular, these operations have risks of causing inner ear damage and subsequent permanent sensorineural hearing loss as well as vestibular dysfunction [6]. Furthermore, because an abnormal facial nerve course frequently accompanies oval window atresia, traditional atresia surgery is complicated and sometimes impossible [5, 6]. The recent work by Colletti et al. [9] reports that "round window vibroplasty" is an option to provide better hearing outcomes relative to traditional vestibulotomy with ossiculoplasty, with lower risks to the patient.

Recent reports suggest that Bonebridge, a novel implantable bone conduction hearing aid, is safe and effective for treating conductive hearing loss. The technique for insertion is easier than that of traditional surgeries, and there is no risk of inner ear damage. Because the internal part of the device can be fully implanted without abutment, there is no risk of skin infection. In addition, the active vibrating part, known as the floating mass transducer (FMT), is also implanted. This ensures that the Bonebridge hearing system provides more bone conduction gains than the gains of passive bone conduction devices [7, 8]. Because FMT is bigger than other devices in the market, making enough bony space for the device can be challenging, particularly in children or patients who underwent previous mastoid surgeries [10].

Regarding this case report, because the absence of stapes and the abnormal facial nerve course around the round windows in the patient

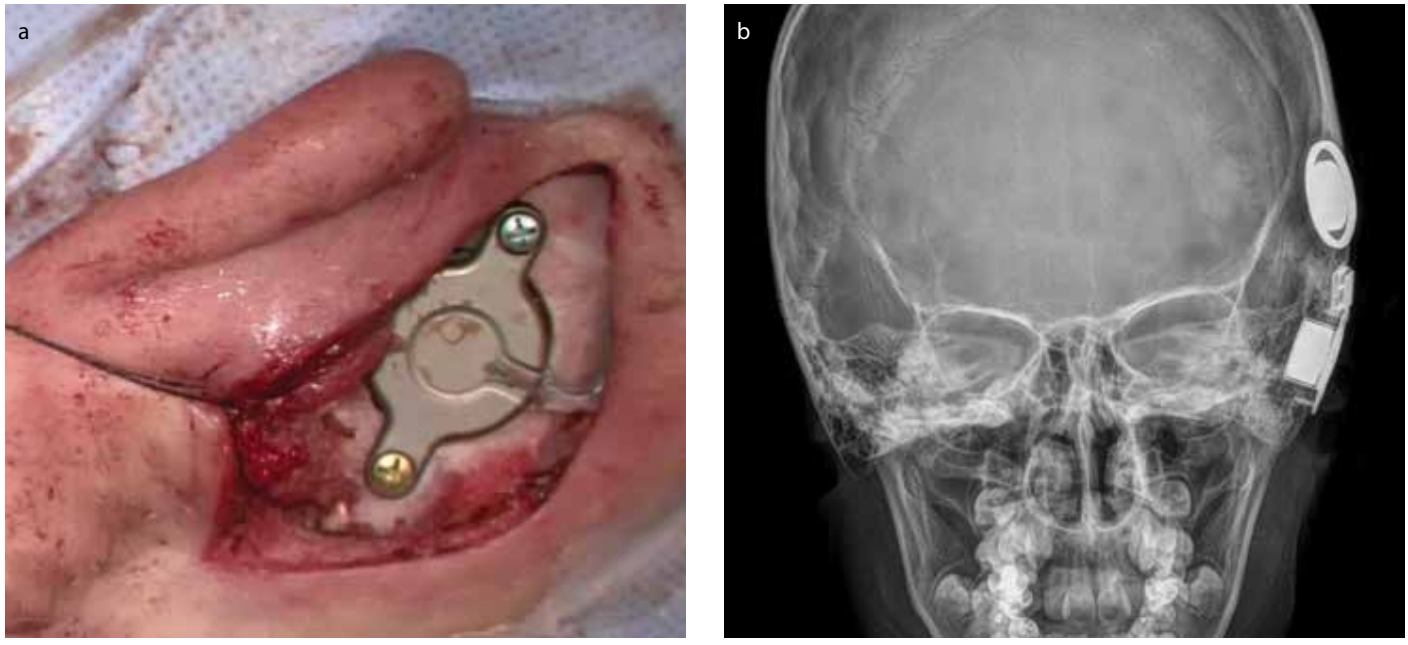

Figure 3. a, b. Surgical findings and postoperative X-ray. The internal device was fixed at the left mastoid area (a). Transorbital view revealed the appropriate position of the internal device (b)

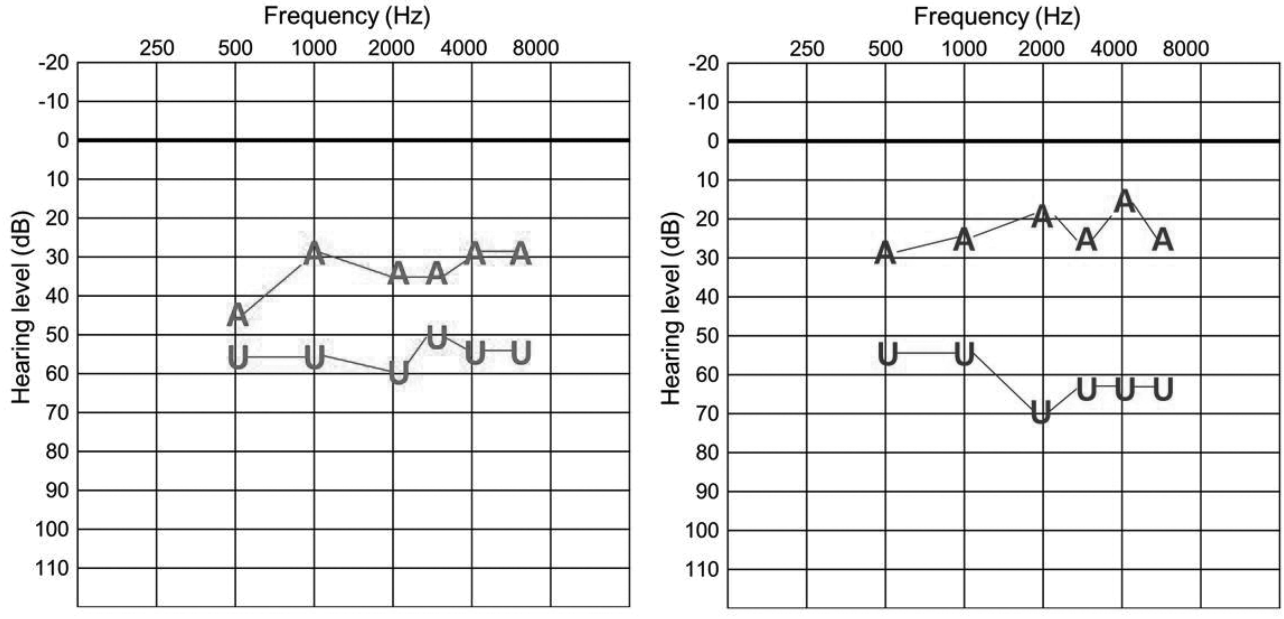

Figure 4. Aided pure tone audiogram in the free-field condition. U: Unaided hearing threshold, A: aided hearing threshold

prevented not only traditional vestibulotomy but also round window vibroplasty, we assumed that Bonebridge implantation was a reasonable solution for her hearing loss. Because Bonebridge is an active transcutaneous bone conduction device in which the external sound processor transmits electromagnetic signals through the skin into the implanted transducer, it can achieve bone conduction hearing gain without the occlusion effect in patients with oval window atresia [11].

In terms of the postoperative hearing of the patient, we observed dramatic improvement in speech audiometry, as well as in field pure tone audiometry. The patient's SRT decreased from 66 dB to 18 dB after the surgery, and SDS increased from 92% at MCL of 90 dB to 100% at MCL of 50 dB. In addition, the patient was more satisfied with the better sound quality from Bonebridge than the sound quality from other conventional hearing aids. Because there is no standard method to evaluate the subjective improvement of sound quality from this device, it is necessary to develop a questionnaire to score the improvement of sound quality of Bonebridge in comparison to conventional hearing aids [7].

For the treatment of conductive hearing loss in patients with oval window atresia, Bonebridge implantation is a promising option with-

out the challenges of difficult surgical techniques or the risks of inner ear damage.

Informed Consent: No written informed consent was obtained as this was a case report preserving anonymity without recognizable photographs of the patient.

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