

# Simultaneous vibrant soundbridge implantation and 2<sup>nd</sup> stage auricular reconstruction for microtia with aural atresia

Lynne Hsueh Yee Lim, Ling Xiang, Jocelyne del Prado, Ee Ling Chin, Millo Achille Beltrame

Otolaryngology – Head & Neck Surgery, National University Hospital, Singapore

## Abstract

Aural atresia and severe microtia are associated malformations that result in problems with hearing and cosmesis, associated speech and language difficulties and diminished self-esteem. In cases where middle ear ossiculoplasty and aural atresia canalplasty are expected to give poor hearing outcomes that would eventually require the use of hearing aids, bone anchored hearing aids or active middle ear implants may be better options. This case report describes a simultaneous Vibrant Soundbridge implantation and 2<sup>nd</sup> stage auricular reconstruction with rib graft cartilage for an 11-year-old boy with grade III microtia and aural atresia 8 months after the 1<sup>st</sup> stage reconstruction. Audiometric results of the Vibrant Soundbridge aided ear were comparable to that of the contralateral hearing aid aided ear.

## Introduction

Aural atresia and severe microtia are associated malformations in many patients due to the common embryologic origin. They can result in problems with hearing and cosmesis, associated speech and language difficulties and diminished self-esteem. In cases where middle ear ossiculoplasty and aural atresia canalplasty are expected to give poor hearing outcomes that would eventually require the use of hearing aids (HA), bone anchored hearing aids (BAHA) or active middle

ear implants (MEI) may be better options.<sup>1-7</sup> A BAHA transmits sound through the skull to both inner ears with almost the same energy, thus speech perception and directional hearing are poorer in unilateral atresia patients aided with BAHA compared to those with normal hearing.<sup>5</sup> The need for daily maintenance of the percutaneous pedestal and the increased risk of recurrent local inflammation are the main concerns for BAHA.<sup>5</sup> Vibrant Soundbridge (VSB) is a semi-implantable MEI with the external processor held onto the head with a magnet, thus eliminating the problems with maintenance and local infection. It was originally intended for patients with moderate to severe sensorineural hearing loss. Since Colletti *et al.* placed the floating mass transducer (FMT) directly on the round window for 7 patients with mixed hearing loss and obtained good results, successful attempts have been made to position the FMT onto various middle ear structures.<sup>8-11</sup> This allows the VSB to be used in cases of conductive and mixed hearing loss successfully. Simultaneous surgery of middle ear implantation and auricular reconstruction has been rarely reported worldwide.<sup>5-7</sup> When successfully done, however, it cuts down the cumulative surgery time, hospital stay, and rehabilitation period for the patient.

## Case Report

An 11-year-old boy presented with bilateral moderate-severe conductive hearing loss with grade III microtia and aural atresia in the right ear. The left ear had a stenotic ear canal but a normal auricle. The right ear canal atresia precluded the use of HA, so he had been wearing a left HA (Siemens Prisma) since young. However, the left HA was complicated by ear canal infections requiring frequent medical treatment, thus the use of HA was intermittent. His mother said he was struggling in school with poor academic grades and performance. He was also very reserved and talked little due to poor self-esteem from his microtia cosmetic concerns. The pure tone audiometry (PTA) showed moderate-severe conductive hearing loss in both his ears. The mean threshold (refers to the mean air conduction threshold, calculated as the average of thresholds at 500, 1K, 2K, and 4K Hz in this case report) was 66 dB HL in the right ear and 60 dB HL in the left ear (Figure 1). Bone conduction thresholds were normal on both sides. Aided speech discrimination test was performed on the left ear using the CID W-22 list, which was presented at 40 dB HL (approximately 60 dB SPL) through a speaker at 90 degree azimuth and 1 meter away from the patient. Noise was presented at 30 dB HL (10 dB signal-to-noise ratio) through the same speaker. The patient scored 20/25 (80%) and 19/25 (76%) respectively in quiet and in noise with the left HA, suggesting that he had slight difficulty discriminating monosyllabic words. The patient had not been tested for language ability before as there were no normed language assessments available in his mother language (Filipino). The patient's language ability was assessed informally by obtaining a language sample through the

Correspondence: Lynne, Hsueh Yee Lim, 1E, Kent Ridge Road, NUHS Tower Block, Level 7, Otolaryngology Head & Neck Surgery, Singapore, 119228. Tel: +65.67725308 - Fax: +65.67753820. E-mail: enthyl@nus.edu.sg

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conversation with him. His speech was assessed using the GOS.SPASS to highlight specific speech sounds that he would be missing out. He was found to have severe delay in expressive language development, which would also impact his academic performance. He was also found to have weak sound discrimination skills which affected his ability to remember long complex instructions that were 2 steps or more. He could not differentiate well between words that sound similar. He refused his front and back sounds (e.g. /k/->/d/, /g/->/t/), the manner of production (e.g. /v/->/b/, /s/->/t/), and occasionally voiced and voiceless sounds (e.g. /t/->/d/). His speech production was inappropriate according to his chronological age. Cognitive test was not performed as the parents were not keen on it. Low middle ear compliance (compliance = 0.2 cm<sup>3</sup>) and elevated negative pressure was observed in the left ear on tympanometry suggestive of abnormal middle ear function. CT scan of the temporal bone showed an atretic external auditory canal and abnormal middle ear structure in the right ear. The facial nerve's course was significantly more anterolateral than usual (Figure 2). Ossiculoplasty and atresia canalplasty in the right ear were not performed as the patient's Jahrsdoerfer score was only 6 (0 score for the middle ear space, facial nerve, malleus-incus complex,

and appearance external ear).<sup>12</sup> The family and patient gave the reason of not wanting a screw exposed on the scalp when asked why they chose the VSB over BAHA. And for the similar reasons, their preference was for cosmetic reconstruction using rib graft cartilage instead of a prosthetic ear mounted over implanted titanium screws on the skull. Thus the 1<sup>st</sup> stage right microtia reconstruction with rib graft cartilage was performed. During the surgery, the patient's own rib grafts were harvested and fashioned into a 3-D framework, and then inserted into the skin flap pocket around a vascular pedicle. The 2<sup>nd</sup> stage auricular reconstruction together with the VSB implantation was performed simultaneously 8 months later. The auricular framework was raised first. An anterolateral mastoidectomy approach was used, keeping high (just under the tegmen) and anterior (just behind the temporo-mandibular joint) to avoid the anomalous facial nerve. A well was drilled posterior to the mastoid cavity to house the fully recessed VSB device. The handle of malleus was absent. The remnant head of malleus was fused to the head of incus. The incus and stapes were otherwise normal. The ossicular chain was mobile after the atretic plate removed and epitympanum and middle ear adhesions freed. The FMT was clipped onto the abnormal malleus-incus

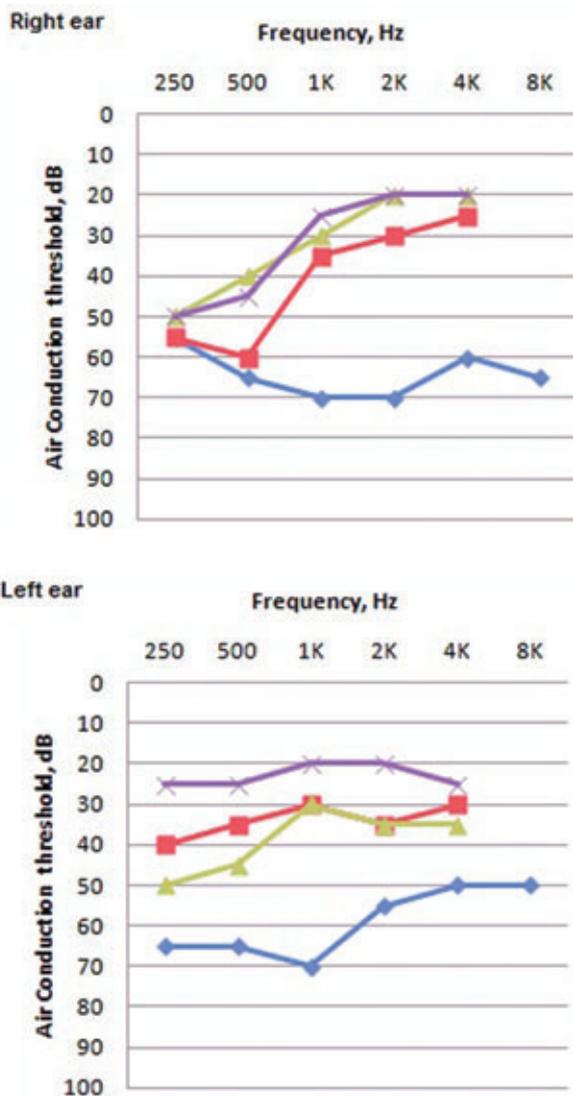


Figure 1. PTA of the right (VSB aided) and left (HA aided) ear. Diamond = unaided before implantation; square = aided 6 months after VSB activation; triangle = aided 12 months after VSB activation; cross = aided 18 months after VSB activation.

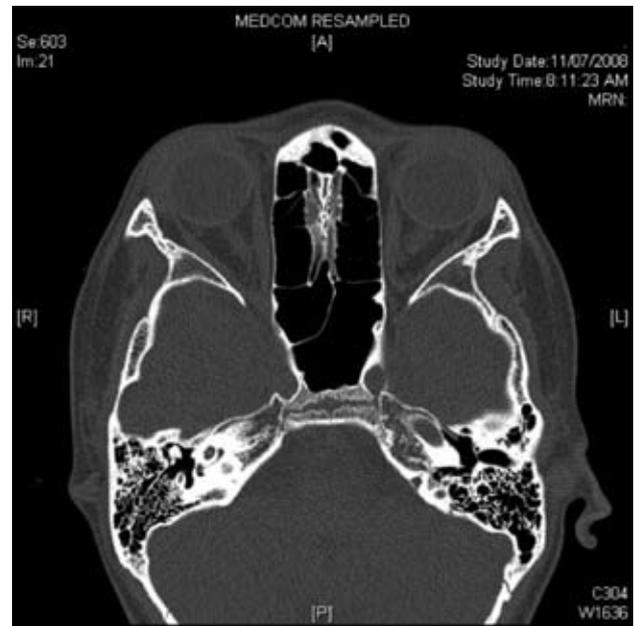


Figure 2. Axial CT image of the temporal bone.

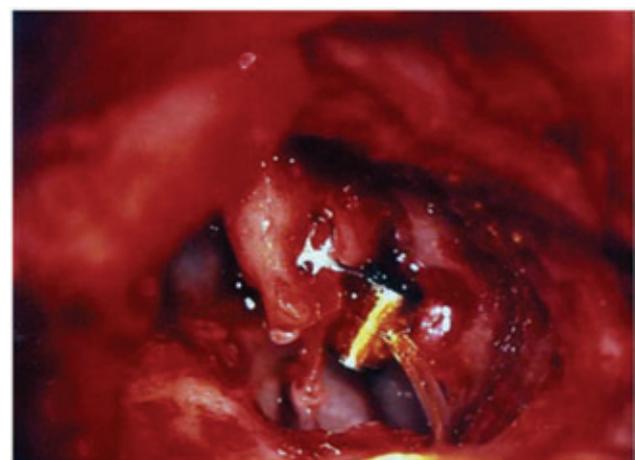


Figure 3. The FMT was clipped onto the abnormal malleus-incus complex in the patient's middle ear.

complex (Figure 3).

For improved vascularisation of the 3-D rib graft auricular framework, a temporoparietal fascia flap based on the superficial temporal artery was raised. The cartilage crescent was inset over the mastoid to provide sulcus and projection of auricle. The temporoparietal flap with superficial temporal artery was then turned down and inset onto the back of the raised auricular framework. Split skin graft was harvested from the abdomen and used to cover the back of auricle and part of mastoid. There was pressure necrosis alopecia on the left scalp due to the long surgery of 8 hours (simultaneous auricular reconstruction and VSB implantation). However, the hair growth returned over that site 5 months later. There were no complications of facial palsy or cerebrospinal fluid leak. The patient was discharged after 2 days. The healing of the auricle reconstruction was uneventful.

The VSB device was switched on 2 months after the surgery. PTA tests were performed at 3, 6, 12, and 18 months post VSB activation on this patient (Figure 1). Results at 3-month follow-up were not reported as the patient's responses were inconsistent. The reliability of responses at the following follow-up sessions was otherwise good. For the right ear, the mean VSB aided threshold was 38 dB HL at 6-month follow-up. It improved to 28 dB HL at 12-month follow-up and remained stable at 28 dB HL at 18-month follow-up. For the left ear, the mean HA aided threshold was 33 and 36 dB HL respectively at 6-, and 12-month follow-up. It improved to 23 dB HL at 18-month follow-up with a new HA (Widex FL-m) fitted. The new HA use was consistent thereafter. The mean unaided bone conduction threshold was 34 and 21 dB HL on the right and left side respectively at 18-month follow-up.

Aided speech discrimination test was performed at 12- and 18-month follow-up. At 12-month follow-up, the patient scored 21/25 (84%) with VSB only and 23/25 (92%) with both VSB and HA (binaural fitting) in quiet. At 18-month follow-up, he scored the same 19/25 (76%) with binaural fitting both in quiet and in noise. The patient reported balanced hearing with both VSB and HA.

Language assessment was not performed postoperatively for two reasons: i) There were no normed language tests in this patient's mother language (as stated earlier); ii) Both the patient's family and school teachers declined the language test as the patient had been doing quite well after the surgery (top 3 student).

The patient's parents completed the hearing device satisfaction scale (HDSS) questionnaire for him at 12-month follow-up. The score was 1.37 (0 = very satisfied; 1 = satisfied; 2 = sometimes satisfied/dissatisfied; 3 = dissatisfied; 4 = very dissatisfied). They also reported improved hearing of the patient in class with the right VSB and the left HA together. This may be one of the causes for the improved school work of this patient. He was also more confident and participating more socially at home and in school.

## Discussion

Surgery for aural atresia combined with microtia has been regarded as one of the most challenging procedures despite significant progress in the field.<sup>13</sup> In 1992, Jahrsdoerfer et al. developed a grading system to measure patients' candidacy pre-operatively, allowing for reasonable prediction of the degree of success of the operation and avoiding operating on impossible cases.<sup>12</sup> Patients with a Jahrsdoerfer score of 7 or higher were found to have significantly better hearing than those with score of 6 or lower.<sup>14</sup> Since our patient's Jahrsdoerfer score was only 6, ossiculoplasty and atresia canalplasty were not performed as the likelihood of good hearing post surgery was low.<sup>12,14-15</sup> BAHA or MEI might be better option for him.<sup>1-7</sup>

VSB has also been successfully applied to patients with microtia and

aural atresia solely or in combination with other surgical procedures.<sup>3-7</sup> In our case, the VSB implantation was integrated in the 2<sup>nd</sup> stage auricular reconstruction with rib graft cartilage. It allows for rehabilitation of hearing and cosmesis together; minimizes the number of surgeries, cumulative surgery time, and time off from school required for the healing process. No major complication was observed in our case and similar studies.<sup>5-7</sup> The residual hearing of those who received simultaneous VSB implantation and auricular reconstruction remained unchanged postoperatively, thus it is safe to combine the two procedures in patients with microtia and aural atresia.<sup>5-7</sup> Care has to be taken to reduce the risk of alopecia from pressure necrosis for such long surgeries. The poor reliability of our patient at 3-month follow-up was probably due to the following two reasons: i) Lack of motivation and cooperation of the patient. This finding indicates the importance of preoperative counseling, and ii) The patient was not used to the sound presented in the right ear due to lack of auditory stimulation since birth. At 6-month follow-up, the patient's reliability improved as he got more used to the sound. VSB aided thresholds at middle to high frequencies (1K-4K Hz) were within the average speech spectrum; however, the improvements at 250 and 500 Hz were limited. At 12- and 18-month follow-up, the patient's VSB aided hearing at 250 Hz improved slightly, while it improved greatly at 500 Hz with fine tuning of the device based on his subjective feedback and the aided test. His mean VSB aided threshold was further improved to 28 dB HL, comparable to that of the HA aided ear.

Similar results were obtained in the study of Kiefer *et al.*<sup>7</sup> The FMT was coupled to the round window of a 33-year-old patient with bilateral microtia and fibrous atresia. The patients' VSB aided thresholds were between 15 and 30 dB HL at frequencies of 750-6K Hz, with limited improvements at low frequencies after the first fitting. Leuwer *et al.* found that the VSB enable the highest hearing improvements at 1K and 2K Hz, while there's a decline of power at higher and lower frequencies.<sup>16</sup> In the study of Frenzel *et al.*, greater improvements were achieved.<sup>5</sup> In the 7 patients with unilateral atresia-microtia, the FMT was placed on the stapes, ossicular chain, or the round window during the plastic auricular reconstruction. The mean aided threshold across subjects was 24 dB HL 6 months post operation.

Despite the limited improvements of the VSB aided ear at low frequencies, it did not affect the overall performance at the speech discrimination test. At 12-month follow-up, the patient scored slightly lower with VSB only (21/25, 84%) than with both VSB and HA (23/25, 92%), which indicates that the poor hearing at low frequencies can be compensated by the contralateral HA aided ear. At 18-month follow-up, the patient scored the same (19/25, 76%) in quiet as in noise with both VSB and HA. However, this score was relatively low compared to 12-month follow-up result. This could be due to the lack of speech and language therapy after 12-month follow-up as both the patient and his parents did not find much difficulty communicating with each other in their mother language. However the current results indicate that VSB enables good hearing thresholds in patient with aural atresia and congenital abnormal middle ear anatomy.

## Conclusions

The VSB enables good hearing thresholds to be achieved in this patient with aural atresia and congenital abnormal middle ear anatomy with poor Jahrsdoerfer score. Simultaneous VSB implantation with auricular reconstruction can be safely performed, allowing for rehabilitation of hearing and cosmesis together, and minimizing the number of surgeries, cumulative surgery time, and time off from school required for the healing process.

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