Hearing rehabilitation in congenital canal atresia

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ABSTRACT

Introduction: The purpose of this study was to review the results of our patients with congenital canal atresia after implantation of bone-anchored hearing aids (BAHA). The occurrence of complications was also reviewed.

Methods: This was a retrospective analysis of the first 16 patients who had BAHA implantation at Universiti Kebangsaan Malaysia Medical Centre, Malaysia. Audiometric assessment was done preoperatively and postoperatively for each patient using the standard procedure. The surgical procedure was described and its complications discussed.

Results: The 16 patients consisted of 11 male and five female patients. Their mean age was 8.9 years at the time of the surgery. The main indication was bilateral canal atresia. 11 patients had implantation of BAHA performed in two stages, while the other five patients had it as a single-staged procedure. The complications that occurred were failure of osseointegration (one patient), granulation tissue overgrowth into the abutment (two patients) and cellulitis surrounding the abutment (three patients). The average preoperative unaided air conduction threshold was 64.9 dB and the average postoperative aided hearing threshold was 29.7 dB. The overall mean functional gain was 35.2 dB.

Conclusion: BAHA has many advantages over the conventional hearing aid in the form of cosmesis, discomfort and hearing gain. It is a reliable hearing rehabilitation tool with good predictable hearing outcome in patients with bilateral canal atresia, especially those unsuitable for canalplasty. Despite its higher cost and the need for surgical implantation, its use is justifiable in properly selected patients.

Keywords: bilateral canal atresia, bone-anchored hearing aid, canal atresia, congential canal atresia, deafness, hearing loss

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INTRODUCTION

The bone conduction hearing aid is a useful alternative to patients who need hearing rehabilitation but are unable to wear an air conduction hearing aid. It can also be effectively utilised by patients who are not suitable surgical candidates for correction of their deficits. However, the conventional bone conduction hearing aid has several disadvantages in the form of cosmesis, discomfort and sound attenuation caused by the soft-tissue layers between the amplifier-transducer and the skull vault.(1,2) The concept of titanium osseointegration in bone conduction hearing aids was initially introduced by Tjellström in 1977.(3) This principle was based on the concept introduced earlier by Bronemark in Sweden. A bone-anchored hearing aid (BAHA) is defined as a hearing aid with percutaneous transmission of sound vibrations to the skull.(1) The use of BAHA gives superior audiometric results when compared to the conventional bone conduction hearing aid. Percutaneous transmission is 10–15 dB more efficient than transcutaneous transmission; however, BAHA has to be surgically implanted to obtain skin penetrating coupling.(1,4)

METHODS

This was a retrospective analysis of 16 patients who had implantation of BAHA at Universiti Kebangsaan Malaysia Medical Centre, Malaysia, from January 2000 to October 2008. The clinical data collected included the gender, type of ear malformation, syndromic association, age at surgery and duration of follow-up. Other parameters such as indications and complications were also analysed.

Audiometric assessment was conducted using standard procedures in a double-walled soundproof room. For each patient, unaided preoperative air and bone conduction pure tone audiometry (PTA) thresholds at 500 Hz, 1 kHz, 2 kHz and 4 kHz were obtained. The unaided preoperative air and bone conduction thresholds were obtained for each patient. The postoperative BAHA-aided soundfield testing PTA thresholds were also recorded for each patient. The postoperative BAHA-aided thresholds were obtained at the time of sound processor fitting and subsequent thresholds were obtained during follow-up at 3–6-monthly intervals. Averages of the preoperative and postoperative PTA thresholds at 500 Hz, 1 kHz and 2 kHz were used for comparison. The overall improvement was measured by subtracting the postoperative BAHA-aided soundfield PTA

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Fig. 1 Photograph shows the abutment situated posterosuperior to the microtic right pinna and canal atresia.

Fig. 2 Photograph shows the sound processor in place.

threshold from the unaided preoperative air conduction PTA threshold. In all cases, BAHA implantation was performed under general anaesthesia. The implantation can be performed in one or two stages. We advocated a two-staged procedure for children below 12 years of age or for patients with poor skull-bone quality such as post-radiotherapy. On the other hand, single-stage implantation was performed in children above 12 years of age. These recommendations increased the chances of successful osseointegration into the patient's skull.

During the first surgery, a 3–4 mm threaded flange fixture was carefully inserted 55 mm behind and 30 mm above the external ear canal. The skin was closed over the fixture. The second-stage surgery was performed at least three months later to allow osseointegration to take place. In the second surgery, the skin around the fixture was raised and trimmed of subcutaneous fat and hair-bearing follicles. The skin covering the fixture was then punched and the fixture exteriorised. The abutment was then attached to the fixture and the dressing was done with antibiotic-soaked ribbon gauze. In the single-stage implantation, the surgical procedures above were done sequentially in one surgery. The patients were followed up one week later for wound assessment. Then the sound processor was fitted at least one month after fixing the abutment (Figs. 1 & 2).

RESULTS

A total of 16 patients had BAHA implantation within the study period. There were 11 males and five females. Their age at implantation ranged from three to 21 years, with a mean age of 8.9 years. 13 patients had bilateral microtia and canal atresia. The other three patients had unilateral microtia and canal atresia with contralateral canal stenosis. Five patients had syndromic association (Table I). All of our patients had used conventional bone conduction hearing aid prior to surgery. Computed tomography (CT) was done in all patients to exclude canal cholesteatoma and to assess the middle ear cavity, including ossicular abnormalities and inner ear structures. All the patients in this series had poor pneumatisation of the middle ear and mastoid with abnormal ossicles, which made them unsuitable candidates for canalplasty. None of them had canal cholesteatoma.

The implantation of BAHA was done in two stages in 11 (68.8%) patients, and in a single stage for the other five (31.3%) patients. The latter five patients were aged above 12 years, with a mean age of 16.2 years at the time of surgery. All procedures were done under general anaesthesia. For the two-stage procedure, the mean time interval between the first and second procedures was 4.1 (range 3–6) months.

In this series, patients were followed up between 4 and 84 months postoperatively, with a mean follow-up period of 54.4 months. One patient with Pierre Robin syndrome had failure of osseointegration. He was six years of age at surgery and the implantation was done in two stages at three months apart. The fixture and abutment were extruded four months after the second surgery. He underwent three revision surgeries for implantation of the titanium fixture within a period of six months before osseointegration was finally successful. Two patients had recurrent overgrowth of granulation tissue into the abutment; this was successfully treated by surgical removal of the granulation tissue and laying a split-skin graft over the exposed periosteum. Another three patients had cellulitis surrounding the abutment, which had resolved with oral and topical antibiotics.

The average preoperative unaided air conduction threshold was 64.9 (range 47–73) dB (Table II). All patients had a preoperative bone conduction threshold of below 20 dB. The average postoperative BAHA-aided soundfield hearing threshold was 29.7 (range 18–42) dB. There were no changes of the postoperative BAHA-aided soundfield

Table I. Demography, surgical indications for bone-anchored hearing aid, syndromic association and complications.

Case no.	Age (years)	Gender	Indication for surgery	Syndromic association	Surgical procedure (stages)	Complications	Management
1	7	F	Bil CA	Goldenhar	2	–	Reimplantation with sleeper
2	6	M	Bil CA	Pierre Robin	2	Extruded fixture × 2	–
3	5	M	Bil CA	–	2	–	–
4	14	M	Bil CA	–	1	–	–
5	5	M	Bil CA	–	2	Cellulitis around abutment	Conservative
6	16	M	Bil CA	Treacher Collins	1	Recurrent granulation tissue around abutment	Excision of granulation tissue & split-skin grafting
7	15	F	Bil CA	–	1	–	–
8	6	M	Bil CA	–	2	Cellulitis around abutment	Conservative
9	5	F	Bil CA	–	2	–	–
10	7	F	Right CA & left CS	–	2	–	–
11	3	M	Bil CA	Townes-Brocks	2	–	–
12	15	M	Bil CA	–	1	–	–
13	9	F	Left CA & right CS	–	2	Cellulitis around abutment	Conservative
14	3	M	Left CA & right CS	1st & 2nd branchial arch syndrome	2	Granulation tissue around abutment	Excision of granulation tissue & split-skin grafting
15	5	M	Bil CA	–	2	–	–
16	21	M	Bil CA	–	1	–	–

Bil: bilateral; CA: canal atresia; CS: canal stenosis

hearing thresholds obtained at the time of sound processor fitting or during subsequent follow-up. The overall mean functional gain was 35.2 (range 23–55) dB. At the time of reporting, all of our patients are using their BAHA and expressed an overall subjective satisfaction with their hearing outcome.

DISCUSSION

Congenital abnormalities of the middle and external ear are seen more often than in the inner ear. In the case of unilateral canal atresia, acquisition of speech and language is possible provided that the patient has normal hearing thresholds in the normal ear. However, in bilateral canal atresia, hearing amplification with bone conduction hearing aid should be initiated as early as possible to enable normal speech development. This can be achieved by means of a bone conduction hearing aid at the infant stage. We advocate the use of bone conduction hearing aid as early as three months of age in those who have bilateral canal atresia. Currently, an infant can wear a sound processor attached to a soft band, which is convenient and comfortable.

The surgical options available for patients with canal atresia include canalplasty and BAHA. Canalplasty can be technically difficult with variable outcomes. There is also a risk of facial nerve injury, canal restenosis and chronic otorrhoea. Postoperative management is challenging and more so if the child is uncooperative. In our centre, high-resolution CT (HRCT) of the temporal bone is performed in patients with canal atresia at the age of four years. The main aim of the imaging studies is to exclude canal cholesteatoma, to assess the degree of pneumatisation of the middle ear and mastoid, and to assess the middle ear structures (ossicles and facial nerve) and inner ear structures (cochlea). Surgical exploration is indicated if canal cholesteatoma is present. In addition, canalplasty is recommended if there is almost normal pneumatisation of the middle ear and mastoid and normal placement of the facial nerve, with the presence of normal or near normal ossicles and inner ear structures. This is because the main aim of canalplasty is to achieve a functional hearing outcome. On the other hand, if the middle ear contents and pneumatisation are abnormal, we recommend BAHA as an alternative hearing tool in these selected cases because of its favourable hearing outcome and low complication rate.

In general, the indications for BAHA are divided

Table II. The hearing outcomes of patients who underwent implantation of the bone-anchored hearing aid.

Case no.	Age at surgery (years)	Surgical procedure (no. of stages)	Preoperative air conduction threshold (dB)	Postoperative BAHA-aided soundfield threshold (dB)	Gain (dB)
1	7	2	67	32	35
2	6	2	60	35	25
3	5	2	67	30	37
4	14	1	67	42	25
5	5	2	*	28	*
6	16	1	73	18	55
7	15	1	58	30	28
8	6	2	63	30	33
9	5	2	65	33	32
10	7	2	47	20	27
11	3	2	67	28	39
12	15	1	65	42	23
13	9	2	62	23	35
14	3	2	70	20	50
15	5	2	72	35	37
16	21	1	70	27	43
Mean†			64.9	29.7	35.2

*data unavailable
† of 15 patients

into otological and audiological indications. Otological indications include patients with congenital malformations of the external ear and middle ear canals, acquired canal stenosis, otosclerosis (who are unfit surgical candidates) and external canal closure post skull-base surgery, as well as those with chronically discharging ear or recurrent otitis externa.[2,5] Audiological indications comprise those with conductive or mixed hearing loss and those with single-sided sensorineural deafness. The pure tone average bone conduction threshold of the operated ear should be better than or equal to a 45 dB hearing level, which indicates good cochlear reserve. In our clinical setting, we advocate BAHA implantation in cases of bilateral canal atresia where radiological imaging showed poorly-pneumatised middle ear cavity or abnormal ossicles, because these patients will have poor hearing outcome with canal reconstruction surgery.

The timing of BAHA implantation is dependent on the child's age, although the presence of craniofacial abnormalities should also be taken into account. Previously, the operation was done at the minimal age of five years. At this age, the skull has achieved appropriate skull thickness of at least 4 mm, which is the minimum fixture length used. However, a recent study by Dutt et al showed that the earliest possible age for implant fixture is at two years.[6] As the thickness of the temporal bone is critical for implant integration, many authors suggest HRCT of the temporal bone to assess the skull thickness before planning for fixture implant at two or three years of age.[3] The thinner skull will limit the length of titanium fixture that can be safely placed. Papsin et al reported that a shorter fixture length is associated with a higher failure rate of osseointegration.[7] Concerns were raised with regard to the impact of bony skull growth towards the implant, whereby the implant may become deeply positioned with time. The first child who was implanted by the Gothenburg team was a 13-year-old with mandibulofacial dystosis and the same implant was still being used after more than 20 years.[3]

The implantation of BAHA may be done as a single- or two-staged procedure under local or general anaesthesia. In children and patients with poor bone quality such as post-radiotherapy, the implantation is generally done in two stages. This is to ensure osseointegration of the fixture implant before adaptation of the titanium abutment and skin penetration preparation at 3–4 months. This period may be extended based on the bone thickness and softness, malformation of the temporal bone and the primary stability of the implant. Subsequently, 2–3 months later, the patients are fitted with the sound processor.[3,6] Some authors even advocated the implantation of sleepers that is more than one titanium fixture implant in cases of probable failed osseointegration.[7,8] On the other hand, in teenagers or adults, the BAHA can be implanted in a single-staged operation. A study done by Mylanus and Cremers showed that 94% of their implants were successfully retained in the skull with the longest follow-up period of 25 months.[9]

In our patients, we advocated the single-staged procedure in patients above 12 years of age, because older patients can better care for the surgical wound and are unlikely to have falls that can disrupt fixture osseointegration. In contrast, another centre in Toronto advocated the single-stage procedure in children above five years of age. They concluded that the two-staged BAHA procedure with a prolonged interval between the stages in younger children resulted in hearing outcomes and rates of device failure comparable to those of older children.[10]

The hearing outcome after the BAHA implantation is dependent largely on the preoperative bone conduction PTA thresholds. Almost 90% of patients with bone conduction PTA thresholds below 45 dB, indicating good cochlear reserve, reported an improvement in hearing.[10] In our series, all 16 patients had hearing improvement with a mean functional gain of 35.2 dB, and all reported subjective hearing improvement and are currently using the BAHA device every day. A larger study of 40 patients using BAHA showed that 32 (80%) patients had closure of the air bone gap to within 10 dB of the preoperative bone conduction thresholds.[2] Another recent study done in Glasgow reported that 71% of patients with BAHA experienced an improved quality of life.[11]

The risks associated with BAHA implantation are mainly failure of osseointegration, local skin infection and overgrowth into the abutment.[12] There has been no previous report of osteomyelitis. In our series of 16 patients, one patient had failure of osseointegration requiring three revision surgeries before it was successful. This patient, who had Pierre Robin syndrome, denied any history of trauma. We postulated the possibility of a poorer bone quality in this patient; however, there was no evidence to support our postulation. Another two patients had recurrent overgrowth of granulation tissue into the abutment due to poor hygiene. Finally, removal of the granulation tissue and laying of a split-skin graft over the exposed periosteum proved successful. Another three patients had cellulitis surrounding the abutment which resolved with medication. Therefore, we report an overall 20% (3/15) complication rate (requiring revision surgery), and 6.7% (1/15) implant failure rate. An earlier study of 100 children with osseointegrated implants for BAHA or auricular prostheses reported a 5.8% implant failure rate.[3]

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