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Review

Contemporary solutions for patients with microtia and congenital aural atresia – Hong Kong experience

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Abstract

Managing microtia patients is always a challenge. Multidisciplinary approach, good family support, well established doctor–patient relationship and well organised patient-support groups are the essential elements for success. With the advancement of implantable hearing devices, more options will be available for the microtia patients. Otologists play a leading role in the whole management process. They not only provide proper guidance to the patients in choosing the correct path of the treatment, but also play a key role in organising and maintaining a cost-effective multidisciplinary rehabilitation team for the microtia patients.

Keywords: Microtia; Congenital aural atresia; Implantable hearing aids; Bone conduction

Contents

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1. Introduction

Microtia is a congenital disease of the external ear. Unilateral microtia is more common. The right side is affected almost twice as often as the left and bilateral involvement accounts for 10% of the cases. Some of the microtia patients also have canal stenosis, atresia and/or middle ear deformities.

The incidence of microtia falls within the range of around 3–10 in 10,000 live births (El-Begermy et al., 2009).

Most of the cases are sporadic, however, some form part of the disease spectrum of well known syndromes such as Goldenhar and Treacher Collins (Kelley and Scholes, 2007).

2. Classification of microtia

Microtia patients have malformed external ears of various degrees. There are quite a number of classification systems.

Simply, microtia can be classified as below:

Grade I

- All anatomic subunits are present but abnormally shaped.
- The ear is smaller.
- With or without canal malformation.

Grade II

- The anatomic subunits are deficient or absent.
- The helix is malformed.
- The ear lobe is present.
- Usually with external ear canal malformation.

Grade III

- Classic form of microtia (peanut ear).
- The upper half of the pinna is formed by disorganised cartilage with a malformed earlobe at the lower part.
- Canal atresia is common.

Grade IV

- Anotia.

3. Problems with microtia and canal atresia patients

Microtia patients have 2 main problems:

A) Functional

1. Malformed auricles may make it difficult for the patients to put on glasses or hearing aids.
2. In cases with canal stenosis, the patients may have recurrent wax impaction, recurrent external ear infection and hearing impairment.
3. In cases with canal atresia and/or middle ear deformity, the patients will have conductive hearing loss.

B) Psychological

Malformed external ears may cause significant self-esteem problems in both female and male patients. The problem may be more obvious during teenage. These patients may have difficulties in making friends.

In addition, hearing loss may cause learning difficulties and poor sound localisation. Daily social life and school life is largely affected. These may account for a number of microtia patients have lower self-esteem and introverted behaviour (Tye-Murray et al., 2012).

4. Management of hearing loss in microtia patients

In paediatric microtia patients with aural atresia, the most common type of hearing loss is conductive hearing loss.

Unilateral cases will be managed differently from bilateral cases. In bilateral cases, hearing loss needs to be addressed as soon as possible in order to avoid affecting the speech and language development in children.

For bilateral cases, the treatment options available in our centre are as follows.

Non-Surgical

1. Headband bone conduction devices.
2. Spectacle bone conduction hearing aids

Surgical

1. Canalplasty.
2. Percutaneous Bone Anchoring Hearing Aids.
3. Transcutaneous Bone Anchoring Hearing Aids (BAHA Attract system).
4. Bonebridge.
5. Vibrant Soundbridge.

5. Non-surgical treatment for hearing loss in microtia patients

1. Headband bone conduction devices

Direct sound transmission through the skull bone to the functioning cochlea is made possible by the headband. Paediatric patients waiting for surgical planning and the patients committed for non-surgical treatment are suitable candidates.

The drawback of this device is that firm compression onto the skull bone is required and this may cause skin irritation, headache and occasionally pressure sores. In addition, the device consists of a sizeable microphone and bone vibrator, which brings aesthetic concerns (Fig. 1a and b).

A modern modification of the headband bone conduction system is the replacement of the metal headband by a soft headband with adjustable tension. It has a better outlook and is better accepted by parents and children. Many patients are willing to wear it for most of the days (Fig. 2a and b).

Fig. 1. a. The headband bone conduction device. b. A patient putting on a headband bone conduction device.

Fig. 2. a. The soft band bone conduction hearing aid distributed by Cochlear. b. The soft band bone conduction hearing system distributed by MED-EL.

2. Spectacle bone conduction hearing aids

The sound transmission mechanism of this is the same as the headband bone conduction hearing aid. A good compression onto the skull bone is needed and is made possible by the arms of the spectacle (Fig. 3a and b).

Aesthetically, the spectacle aid looks much better. Functionally, it can solve both the hearing and the eyesight problems for the patient. Even in patients with normal eyesight, the spectacle aid can act as a camouflage for the bone conduction hearing aid. Spectacle aids therefore minimise the stigmatisation problem. However, the patient needs to have a reasonable external ear remnant size to hold the spectacle. Otherwise, the patient needs to have a prosthetic ear surgery or a pinna reconstruction surgery before the spectacle aid can be fitted.

In addition, spectacle aids are more expensive than the traditional headband bone conduction hearing aids.

6. Surgical treatment for hearing loss in microtia patients

1. Canalplasty

Canalplasty is only indicated in a limited number of patients. The Jahrsdoerfer Classification (Table 1) is a helpful guide during the selection process. Routine audiograms and CT examinations of the temporal bone is required.

Firstly, the patients need to have a normal cochlea reserve. Secondly, canalplasty is only indicated in patients scored 5 or above in our centre.

The baseline of the surgery is to achieve a dry patent self-cleaning canal. In ideal cases, patient's hearing is amplification-free. In general, around 30% of patients are hearing aid dependent even after the canalplasty.

In order to achieve a better success rate, we prefer to operate on older children, usually older than 8 years old, when the children can cooperate for postoperative ear cleaning and dressing.

Facial nerve injury is one of the most common complications encountered and is estimated to be around 1%.

Other complications like post-operative hearing deterioration is around 1% and canal restenosis can be up to around 30% (Teufert and de la Cruz, 2004; Nishizaki et al., 1999).

2. Percutaneous Bone Anchoring Hearing Aid (Percutaneous BAHA)

Percutaneous Bone Anchoring Hearing Aid was introduced for treating patient with conductive hearing loss in the 1970's. The components of the system include a sound processor, an abutment and a titanium fixture (Fig. 4).

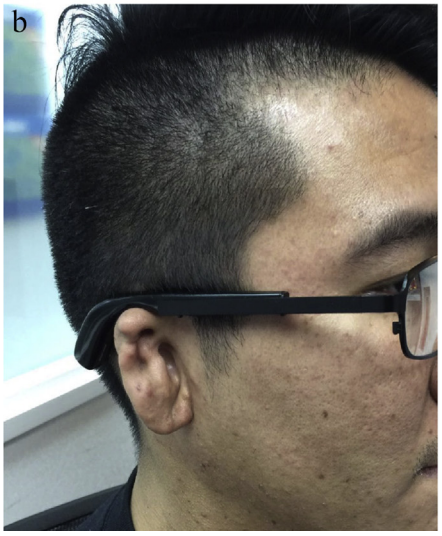


Fig. 3. a. The spectacle bone conduction hearing aid. b. A microtia patient wearing a spectacle bone conduction hearing aid.

Table 1
Jahrsdoerfer classification.

CT temporal bone findings	Score
Stapes present	2
Middle ear space	1
Oval window open	1
Facial nerve normal	1
Malleus-incus complex present	1
Mastoid well-pneumatized	1
Incus-stapes connection	1
Round window normal	1
Appearance of external ear	1

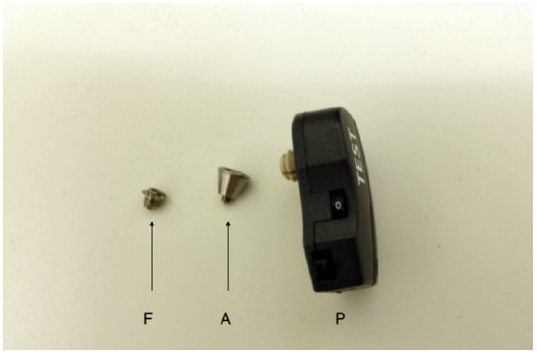
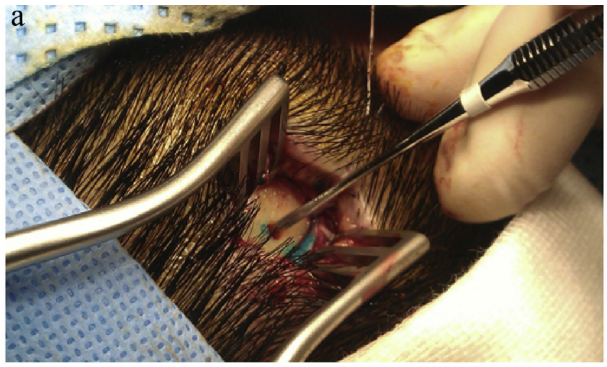


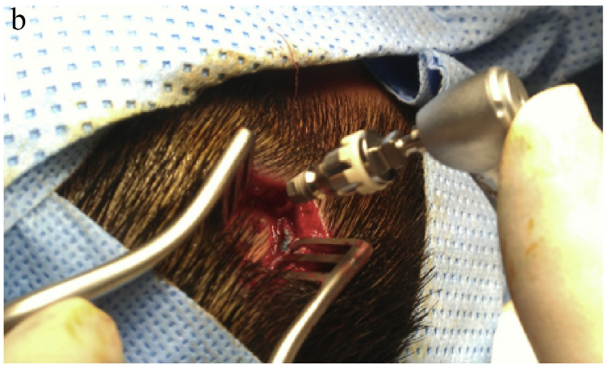
Fig. 4. Components of the percutaneous bone anchoring hearing aid system. The fixture (F), the abutment (A) and the sound processor (P).

The system makes use of the bone conduction and the osseointegration concepts. Osseointegration provides a secure interface for sound transmission from the hearing implant to the functioning cochlea. This is made possible via the implantation of a titanium fixture, which provides a direct connection, both functionally and structurally between the living bone and the implant for sound transmission.

Implantation is by surgery. A small skin incision is made at the temporal area (Fig. 5a). The fixture is implanted and the abutment is connected to the fixture through a small skin puncture made at the scalp (Fig. 5b). A small amount of tissue reduction is made at around the periabutment site (Fig. 5c).

BAHA sound processor fitting can be performed after full osseointegration and complete skin wound healing. This is





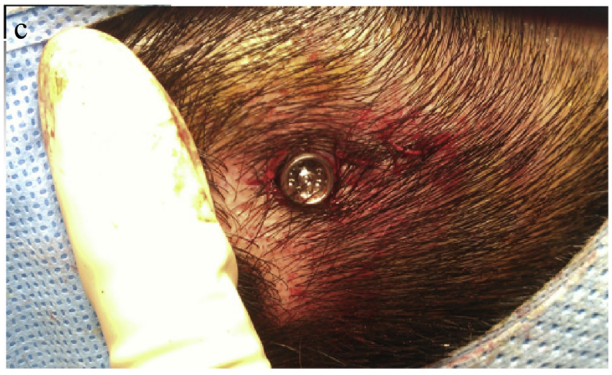


Fig. 5. a. A skin incision was made and a bony well created at the temporal bone area for the accommodation of the BAHA Titanium fixture. b. The Titanium fixture that pre-mounted with an abutment was about to be implanted in the single-staged Percutaneous BAHA Surgery. c. The appearance of the periabutment site after the soft tissue reduction has been carried out.

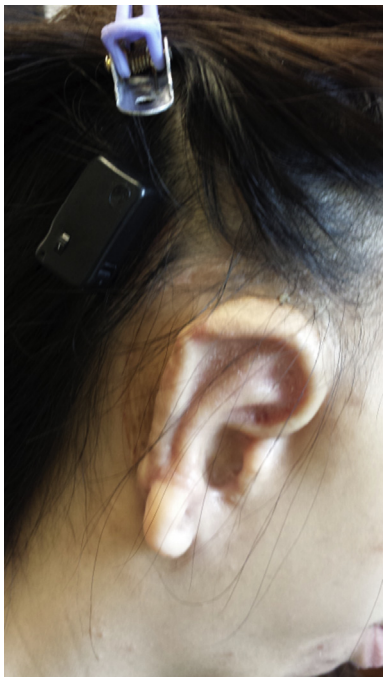


Fig. 6. A right microtia patient fitting on a Percutaneous BAHA. She also had pinna reconstruction performed for her microtia.

usually around 3 months after the surgery (Fig. 6). In patients with thin skull bone, like paediatric patients, a staged procedure is recommended, where the titanium fixture has to be implanted first and another abutment fitting surgery is then carried out 3 months later, so that the osseointegration process can be carried out fully without disturbance.

3. Transcutaneous Bone Anchoring Hearing Aid (BAHA Attract system).

This is a new device by Cochlear (Fig. 7a). Without an open percutaneous pin tract wound, the sound processor is attached to the scalp by magnetic coupling. The surgery is simple. A postaural incision is made for the fitting of a Titanium fixture (Fig. 7b) and the magnet is implanted under the skin (Fig. 7c). The sound processor can be switched on 3–4 weeks after the surgery when the postaural wound has healed completely (Fig. 7d).

The beauty of the Transcutaneous BAHA is that it is without any pin tract wound problems. In our centre, the surgery can be performed together with the second staged pinna reconstruction using a two-team approach.

4. Bonebridge (BB)

Bonebridge is an implantable device by MED-EL, Innsbruck, Austria (Fig. 8a). It is a bone conduction device without pin tract problems. The Bonebridge has two parts, the external Audio Processor and the implanted part, the Bone Conduction Implant (BCI). The surgery needs thorough pre-operative assessment and planning. In our centre, we work with our radiologist colleagues closely. Navigation CT scan and pre-operative accurate measurements are performed for identification of the best implantation site (Law et al., 2015). The mastoid

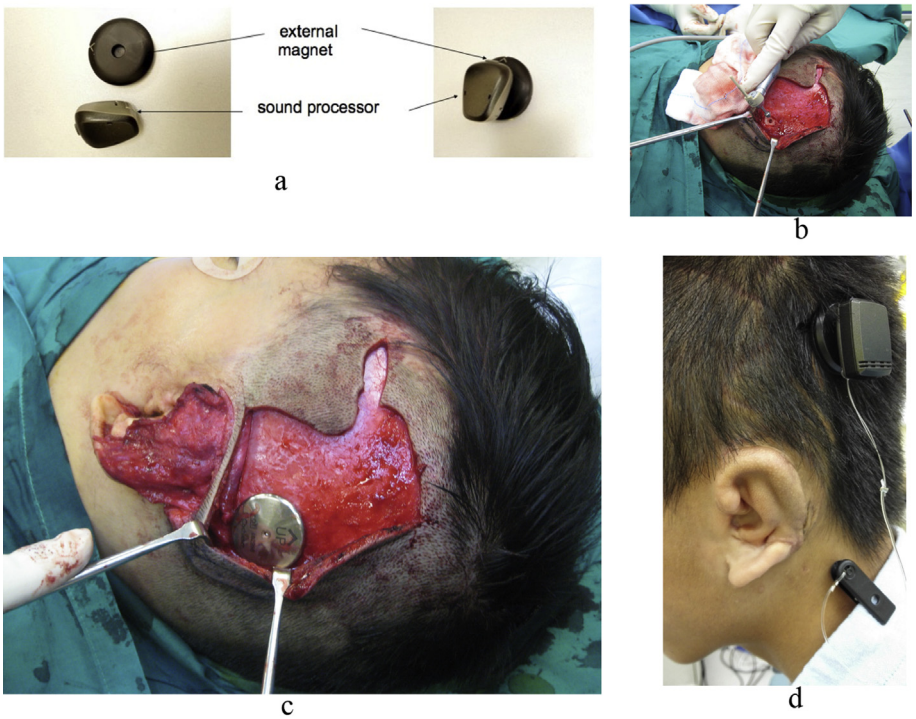


Fig. 7. a. The Transcutaneous Bone Anchoring Hearing Aid (BAHA Attract system) distributed by Cochlear. b. A patient with left Transcutaneous BAHA surgery together with the second-stage pinna reconstruction surgery. A Titanium fixture was about to be implanted at the temporal bone. c. The same patient with the implant magnet (internal magnet) fitted with the BAHA Titanium fixture. The magnet will be covered by the healthy scalp skin upon the completion of the pinna reconstruction surgery. d. The same patient with the BAHA Attract System.

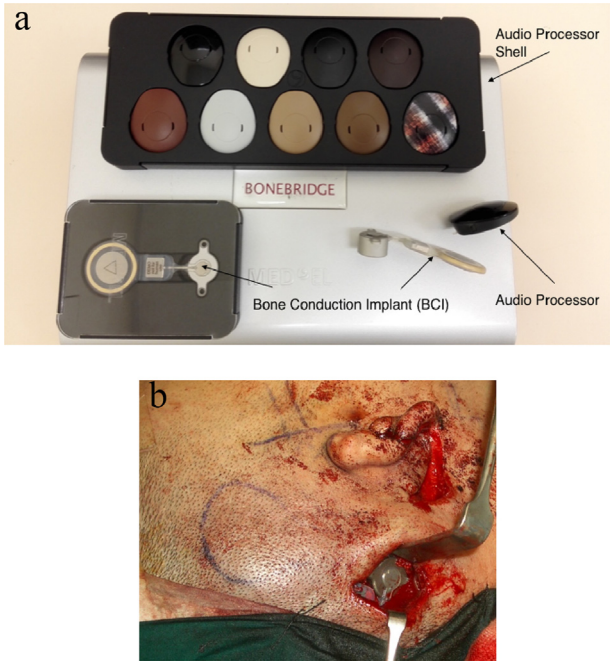


Fig. 8. a. The Bonebridge. b. A right Bonebridge implanted at the mastoid region of a microtia patient.

area is the commonest site for implantation (Fig. 8b). In some cases like patients with contracted mastoids and prominent sigmoid sinuses, a retrosigmoid approach is indicated.

Switching on the device can be done 3–4 weeks after the operation when the surgical wound has healed completely.

5. Vibrant Soundbridge (VSB)

This is a semi-implantable device by MED-EL, Innsbruck, Austria (Fig. 9). It provides a direct stimulation to the inner ear by being implanted at the ossicles or the round window (Vibroplasty).

It is different from bone conduction devices like BAHA and bonebridge because the VSB does not stimulate the cochlea bilaterally and will not result in signal confusion and incorrect sound localisation.

The VSB implant consists of two components, the external audio processor (AP) and an implanted part, the Vibrating

Fig. 9. The Vibrant Soundbridge.

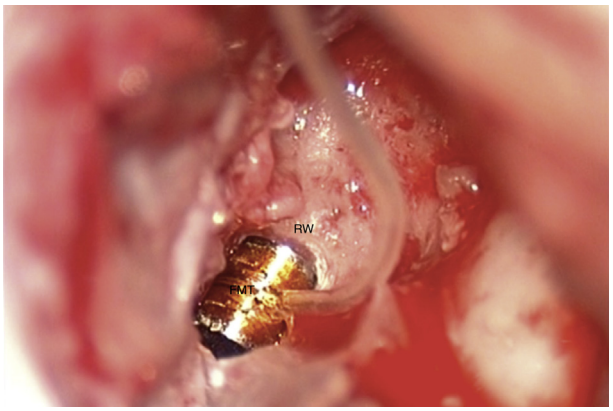


Fig. 10. Left Vibrant Soundbridge Surgery – Vibroplasty Technique. RW = round window, FMT = Floating Mass Transducer.

Ossicular Prosthesis (VORP). A Floating Mass Transducer (FMT) attaches to the distal part of the VORP. The FMT vibrates the attached middle ear structure through a single point attachment and stimulates the inner ear.

In microtia patients with aural atresia, the surgical approaches commonly employed are the round window approach (Vibroplasty) (Fig. 10) and the stapes coupling (Fig. 11).

There are different kinds of couplers to facilitate the best fitting of the FMT onto the ossicles or the round window (Fig. 12a and b).

The device can be switched on 8 weeks after the operation when the surgical wound has healed completely.

7. How to choose a suitable device for the microtia patients with hearing loss?

In our centre, we will consider the following factors when counselling the patients and the family.

1. Unilateral vs Bilateral hearing loss

In bilateral cases, especially in children, treatment of hearing loss is urgent. A headband bone conduction device or a soft

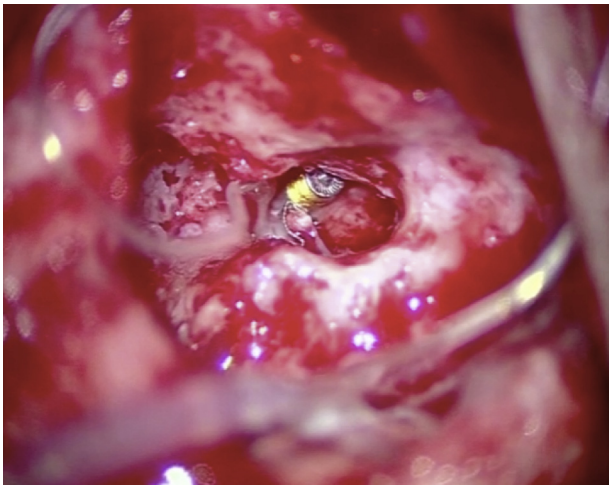


Fig. 11. Right Vibrant Soundbridge surgery – stapes coupling technique.

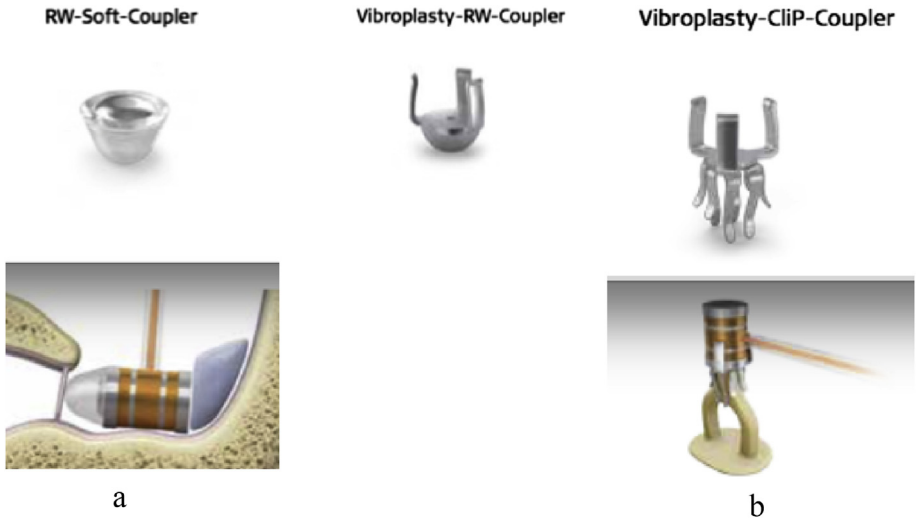


Fig. 12. a. Round window Couplers of the Vibrant Soundbridge System (Courtesy of MED-EL). b. A Vibroplasty-CliP-Coupler of the Vibrant Soundbridge System, for the FMT placement onto the head of the stapes when the stapes is strong enough and mobile (Courtesy of MED-EL).

band bone conduction aid is indicated. The patient will be monitored for speech and language development closely and appropriate intervention will be introduced where indicated.

In unilateral cases, if the patient does not have speech or language developmental delay, we will review the patient regularly and offer thorough discussion of the available intervention options at follow-ups.

2. Age

Non-surgical devices

Neither the headband bone conduction hearing aids nor the soft band bone conduction hearing aids has any age limit for fitting.

The Spectacle bone conduction hearing aids are usually recommended for adults.

Surgically Implanted Hearing Devices for patients

i) Less than 5 years old

Vibrant Soundbridge (VSB) in selected cases.

ii) Older than 5 years old

Vibrant Soundbridge (VSB), Bonebridge (BB), Percutaneous BAHA, Transcutaneous BAHA (BAHA Attract system) are the available options.

Canalplasty and Tympanoplasty

Indicated in selected patients older than 8 and the Jahrsdoerfer Classification is a helpful guide during the selection process.

3. Thickness of the skull bone

Thickness of the skull bone is a determining factor for the selection of specific type of implantable aids.

The BAHA system is advised to be performed on skull bone with thickness of 3 mm or more. The Bonebridge (BB) is even more demanding, as the size of the Bone Conduction Floating Mass Transducer is the largest (8.7 mm × 15.8 mm). However, with the new spacer system – the BCI lift (Fig. 13a), it is possible to have the surgery on thinner skulls (Fig. 13b).

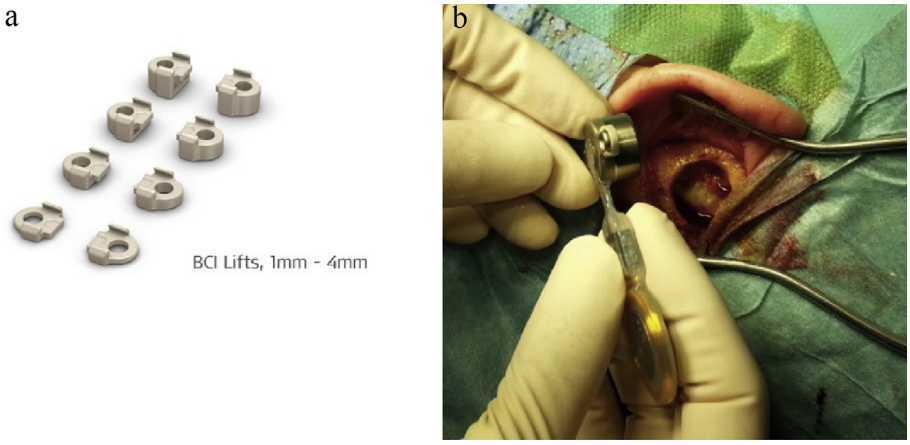


Fig. 13. a. The BCI lifts for the Bonebridge (Courtesy of MED-EL). b. A bonebridge fitted with the BCI lifts.

4. Anatomy

For canalplasty cases, the anatomy of the middle ear structures is the major determining factor for success. The Jahrsdoerfer Classification is a helpful guide during the selection process and good outcome is expected in patients having Jahrsdoerfer Score 7 or above.

High riding Jugular bulbs and abnormal facial nerve positions in the microtia patients will make the Vibroplasty difficult. Whenever there is a round window obliteration, Vibroplasty cannot be chosen.

5. Patient's Medical Health

Some of the implantable devices are MRI conditional, like the Vibrant Soundbridge, the Bonebridge and the BAHA Attract. They can tolerate static magnetic field up to 1.5 T. They also produce artefacts in the images of the head and the brain.

Implantable hearing devices will not be the best option for patients requiring regular MRI follow up of diseases in the head and neck region.

Monopolar diathermy is also contraindicated in implanted patients when the surgery is in the head and neck regions.

6. Family and patient's expectations

In our centre, Canalplasty is carried out in patients aged 8 or above. This is because regular post-operative wound care and aural toileting is required, thus a more mature and highly cooperative patient is more likely to be successful.

Percutaneous BAHA needs the patient's commitment and the support of the family members for a life-long pin tract dressing.

Tuning of the implantable devices to the best fitting level can be lengthy and it may take months after the surgery.

All these post operative care procedures are time consuming and requires the patient's understanding, cooperation and patience. Full engagement of both medical staff and the patient is the key for success.

7. The Rehabilitation Team

A strong multi-disciplinary support is vital to the success in managing hearing loss for the microtia patients.

A good working group of Otologists, Plastic Surgeons, Nursing staff, Social workers, Speech therapist, Radiologists and Audiologists is essential.

Acknowledgements

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