

Paediatric Percutaneous Bone Anchored Hearing Devices

Special considerations in children

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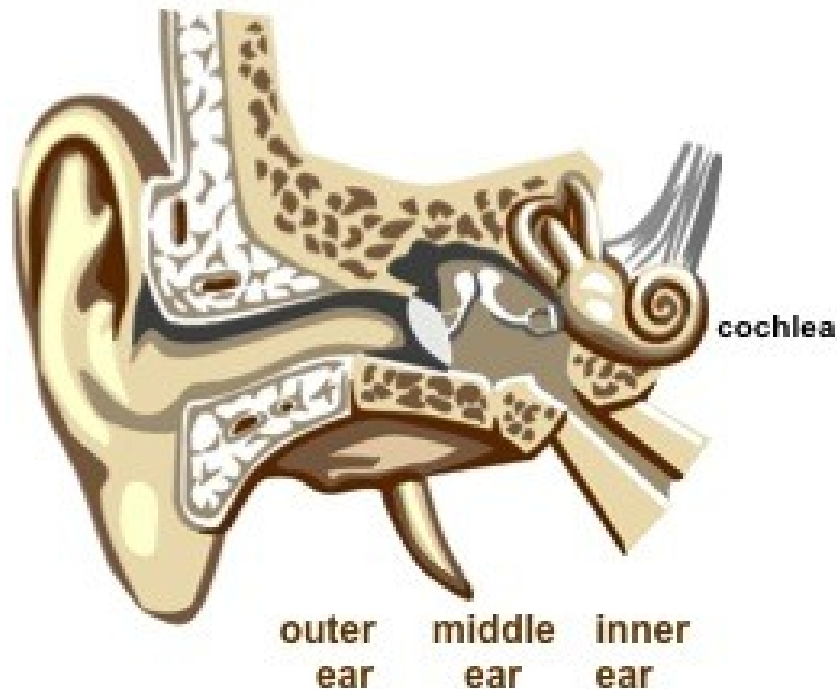
Chapter 1-1

General Introduction

Introduction

Anatomy and physiology of hearing

The acoustic nerve is activated by sound stimulus that can gain access to the nerve via two pathways, air conduction and bone conduction.



Air conduction pathway

In air conduction, the pinna and external ear funnel acoustic signals into the ear and then the ossicles in the middle ear play an important role in matching the impedance of the air filled external ear and the fluid filled cochlea. The sound vibrations cause a travelling wave of fluid within the cochlea. The hair cells located on the basilar membrane are arranged in a tonotopic fashion. The hair cells detect this movement and in turn convert mechanical energy into electrical activity and directly stimulate the acoustic nerve fibres.

Bone conduction pathway

Bone conduction pathways are not as fully understood as the air conduction pathways. The concept of hearing by bone conduction was first popularized in the 1930s by Von Békésy.¹ He discovered that cochlear hair cell excitation was the same irrespective of whether the sound wave pathway was via air or bone conduction. In 1966 Tonndorf described 3 mechanisms by which bone conduction

can stimulate the cochlea: bone compression, inertia bone conduction and the sound energy of the external ear.²

1. Bone compression mechanism

As the vibrations reach the cochlea, the bony cochlear suprastructure vibrates. As the fluid within the cochlea is non compressible, this vibration causes movement of the fluid. As the scala vestibuli (which in turn is connected to the vestibule) is of a larger volume than the scala tympani and as the round window is more compliant than the oval window, there is preferential displacement of fluid into the scala vestibuli. This in turn causes displacement of the basilar membrane. Additionally, as the round window is much more compliant than the oval window³, this further enhances movement of the basilar membrane. More recent research has found that this mechanism is insignificant for bone conduction hearing.⁴

2. Inertia bone conduction mechanism

Vibration of the skull results in the incus and malleus vibrating as a single unit that moves as a pendulum within the skull. This movement displaces the stapes and stimulates the cochlea in a similar fashion to the air conduction pathway.⁵ This is thought to be of most significance at low and mid frequencies.⁴

3. Sound energy of the external ear

When the skull vibrates with bone conduction sound, this extends into the bony and cartilaginous walls of the external auditory canal causing them to deform. This creates vibrations that are transmitted to the tympanic membrane and beyond as in the air conduction pathway.⁶

Two further mechanisms of bone conduction pathway have been suggested by Stenfelt and Goode: pressure transmission from the cerebrospinal fluid, and inertia of the cochlea fluids.⁴

4. Pressure transmission from the cerebrospinal fluid

Static pressure from the cerebrospinal fluids may be transmitted to the cochlea via the cochlear aqueduct, but this is unlikely to be a significant pathway.

5. Inertia of the cochlea fluids

This is now thought to be the most important factor for bone conduction. Temporal bone vibration causes a relative motion between the cochlear fluids and the

cochlear promontory bone. The oval window, round window and vestibular aqueduct keep this inertia going thus stimulating the end organs of the cochlea.

History of bone conduction aids

The earliest references to bone conduction aids are as far back as the 16th Century with rod devices being used to transmit vibrations via the teeth.⁷ The development of the carbon microphone and magnetic receiver in the early 20th century led to the advent of the bone conduction vibrator.⁸ The first modern bone conduction hearing devices were adapted from existing body worn air conduction aids where the receiver was replaced with a vibrating receiver mounted on a tight fitting headband to hold it onto the mastoid bone.

The advent of transistor technology in the 1960s led to smaller spectacle mounted versions that were more cosmetically acceptable.⁹ Despite the reduction in size of traditional bone conduction devices on a headband, there is still evidence that children – in particular male adolescents find the cosmetic appearance unacceptable.^{10,11}

One of the drawbacks with these types of hearing aid is that a proportion of the amplified sound energy is dispersed within the soft tissues, particularly in the high frequencies.¹² An additional problem relates to the need for significant constant pressure to be applied to the temporal bone. Patients find this uncomfortable and variations in pressure and positioning affect efficacy as well as the underlying skin.¹³

Percutaneous Bone Conduction devices and Osseointegration

The challenges faced by traditional bone conduction aids led to the idea of directly coupling the transducer to the skull. Branemark first developed the concept of osseointegration of a titanium metal screw in bone.¹⁴ He had been doing research on blood flow in the rabbit using a titanium inspection chamber inserted into bone. These chambers needed to be removed in order to re-use them and he discovered much to his annoyance that they were very difficult to remove, thus the discovery of osseointegration.¹⁵

Osseointegration is defined as 'the formation of a direct interface between an implant and bone, without intervening soft tissues'. Initial experimentation on animals with dental implants suggested that the osseointegration was superior if the screws were left unloaded for a period of time.

The first bone anchored hearing device (BAHD) was fitted in 1977 by Tjellstrom.¹⁶

The design of the implant has changed over time in response to further research determining the factors that influence osseointegration.¹⁷ It has been found that the macroscopic and microscopic features of the implant surface can affect osseointegration to improve primary stability and achieve osseointegration faster. Macroscopically an increased diameter and modified threading are thought to improve the stability of the fixture. Microscopically, a roughened surface provides a larger surface area for more direct contact with the bone.

Objective measurement of osseointegration (and hence implant stability) has been a recent topic of discussion as this has implications in the early loading of implants. Currently Resonance Frequency Analysis (RFA) can be used to monitor stability changes over time and there is increasing evidence to support earlier loading of implants.^{18,19}

Cone Beam Computed Tomography (CBCT) has been used in the assessment of intraoral implants. It exposes patients to a lower radiation dose and has recently been used to evaluate temporal bone implants.²⁰

Bone Anchored Hearing Devices in Children – Indications

Initially the concept of the percutaneous BAHD was introduced for those patients with bilateral acquired or congenital mixed or conductive hearing losses who were unable (for practical reasons) to wear a conventional air conduction aid. As the benefit of BAHDs have been evaluated clinically, these indications have expanded over time:

1. Chronic ear disease

Initially bone anchored hearing devices were indicated for patients that had an acquired conductive hearing loss with good bone conduction thresholds caused by chronic ear disease. However the indications have since expanded and reports from the literature suggest that benefit is also derived from those with both sensorineural and mixed hearing losses. In Birmingham the first child was implanted in 1988 and had had previous bilateral mastoid surgery for chronic suppurative otitis media.²¹

2. Congenital abnormality

It was realised very early that the BAHD was an ideal form of hearing rehabilitation for those children with congenital ear abnormalities both with and without a craniofacial diagnosis. The early results from Birmingham reported that the most common indication for a BAHD was congenital aural atresia.²¹ For those children with congenital aural atresia, the options are to reconstruct the external auditory

canal and or pinna, or to provide hearing rehabilitation with a bone anchored hearing device. Evidence suggests that reconstruction surgery may create a cosmetically acceptable auricle, but it does not give acceptable hearing rehabilitation alone.^{22,23}

Currently BAHDs are a safe reliable option that can allow significantly better audiological outcome compared with an unaided reconstruction.^{22,23} Furthermore, they provide a predictable audiological result and do not compromise the option for alternative forms of surgical hearing rehabilitation in future years.

3. Unilateral hearing loss

More recently, the role of BAHD in unilateral hearing losses (of both a conductive and sensorineural nature) is emerging as beneficial.²⁴⁻²⁶

Traditionally it was thought that a child with a unilateral hearing loss and a contralateral normal hearing ear would develop normal speech and language skills and make the same educational progress as a child with binaural normal hearing. However in 2004 Lieu et al²⁷ found that children with a unilateral hearing loss displayed increased grade failure rates, required additional educational assistance and had increased behavioural issues within the classroom. A more recent longitudinal study from Lieu following the progress of children identified with a unilateral hearing loss has shown that although oral language and verbal intelligence quota scores improved with time, school performance did not. In addition to this, both parents and teachers reported persistent behavioural problems and academic weaknesses in about 25% of these children.²⁸ Furthermore, Christensen et al found that in a group of children with profound unilateral sensorineural hearing loss, BAHD implantation improved both hearing in noise testing and scores on the Children's Home Inventory for Listening Difficulties questionnaire.²⁹

It is important to note that not all children with a unilateral hearing loss will go on to have problems. In addition to this, in some cases the aetiology of the hearing loss may be in part the cause of the speech and language delay. The challenge for the clinician is to identify the children that may be helped as early as possible. This is the pathway that is in place in Birmingham regarding the on going care of children with unilateral of bilateral microtia with hearing loss.

Microtia clinic pathway at Birmingham children's hospital

1st appointment for discussion of options

1. *Aiding*

- Bilateral cases BC aiding early (discuss what BC aids).
- Unilateral cases do nothing when they are first referred (however offer detailed discussion to the parent that may want to try aiding earlier).
 - Review at 2 years. If speech delay, glue ear in the good side e.t.c. recommend BC aid.
 - Avoid surgical intervention on the good ear if possible and persevere with BC aid.

2. *Surgical options*

- BAHA age above 4.
- Middle ear implants above age 4.
- Bonebridge (not licenced as yet for children, will be suitable for over the age of 5).

3. *Imaging*

- Request CT temporal bone at age 3-4 with sedation.
- If the patient decides to have vibrant soundbridge or bonebridge will need (baseline) MRI prior to the procedure (at the moment subject to further review).

4. *Atresia surgery*

- No canal atresia surgery offered in this team.
- Ear canal stenosis surgery can be offered.
- Also, there is the option of no action.

5. *Cosmetic reconstruction*

- Prosthetic ear reconstruction at the age of 9 offered by BCH.
- Autologous reconstruction (referral to Great Ormond Street Hospital to be explored).

Further appointments in microtia clinic

Age 2 - for uncomplicated cases (complicated cases i.e. airway problems will be seen as appropriately).

Age 3 - request scans, review speech development and hearing test.

Age 3y 6m - to discuss future intervention or no intervention for auditory implant.
 Age 8 - microtia clinic for cosmetics.

If Family decide for no intervention

Follow up

- a. **Bilateral** by audiology as per audiology protocol.
- b. **Unilateral**
 - If aided, audiology follow up as per audiology protocol.
 - If unaided, see at age 5 (hearing/speech at school) and then at age 7 (cosmetics).
- c. After age 5 follow up every 2 years until age 16.

4. Severely handicapped children

There are reports in the literature supporting the use of BAHD in children with severe learning difficulties and multi-sensory deprivation who have in addition to this a conductive or mixed hearing loss. These children may be unable to undergo the traditional preoperative audiological assessment that is usually performed prior to fitting a BAHD. This makes it difficult to accurately predict benefit. It is vitally important that the clinicians and carers have an open and frank discussion about the aims of any intervention and that the expectations of both parties are realistic. In Birmingham these children are managed with a unilateral BAHD even if there is bilateral hearing loss as this cohort of children have a more difficult postoperative course for both the families and clinicians to manage. Often they do not tolerate appropriate dressings and wound care. Despite this, a positive change in behaviour and increased quality of life noticed by caregivers has been reported.³⁰⁻

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Bone Anchored Hearing Devices in Children – Audiological Considerations

1. Limitations of BAHD in view of BC threshold

Percutaneous and transcutaneous BAHD are limited when patients have a significantly poor bone conduction threshold. Increasing the output of the device can lead to feedback. There are clear fitting ranges available for each of the various devices and their respective processors.

Device	Ave BC dB HL
Oticon Ponto	<45
Oticon Ponto Pro Power	<55
Cochlear Baha 3 (BP100)	<45
Cochlear Baha 3 Power (BP110)	<55
Cochlear Cordell II	<65
Sophono Alpha 2	<45

2. BAHD versus Air Conduction aids with Air-Bone gap

Conventional Bone Conduction Devices have traditionally been considered to have disadvantages over air conduction aids. This is partially due to the discomfort associated with the pressure required for positioning on the mastoid, and also due to the attenuation of high frequencies through the skin and soft tissues. The percutaneous BAHDs have overcome these problems with the direct mechanical coupling of the transducer to an osseointegrated titanium implant.

In patients that have a large air-bone gap, the amplification of an air conduction aid has to be increased in order to overcome the air-bone gap. This increase in gain can then cause feedback. This problem is not encountered with the BAHDs as the performance is not affected by the gap.³²

Bone Anchored Hearing Devices in Children – Challenges

1. Diagnosis

When dealing with a paediatric population there are obvious challenges in the identification of hearing loss and difficulties in testing children at different stages in their development. Many children cannot volunteer a detailed hearing loss history and furthermore they frequently cannot complete the many and often demanding hearing tests possible in the adult population. It is extremely difficult to obtain complex air conduction, bone conduction and masked thresholds in some children. Age appropriate behavioural testing is utilized when possible, but often the younger child or the child with learning difficulties may have to undergo a general anaesthesia in order to obtain hearing thresholds using electric response audiometry.³⁰

Incidence of hearing loss in children:

Although there is some data in the literature regarding the prevalence of permanent childhood hearing impairment and congenital hearing impairment, there is not much data published in the literature regarding the incidence or prevalence of mixed or conductive hearing impairment in children. Furthermore, there is currently no national or international register for hearing impairment.

A recent paper from Baltimore in 2011 estimates the hearing loss in persons of 12 years and older in the United States population by extrapolating data from an epidemiological survey.³³ They found that in this cohort, nearly 1 in 8 has bilateral hearing loss, and nearly 1 in 5 has a unilateral or bilateral hearing loss.

A study from Brazil where a sample of the population were audiotologically tested determined that the incidence of hearing loss (ranging from mild to severe) was 25.9% in persons ranging from less than 4 to the elderly population.³⁴ There was no detail regarding unilateral, bilateral or type of hearing loss.

A further study from London puts the prevalence of deafness at 3.65/1000 at school age, with a larger proportion of neonates being identified at an earlier age due to the introduction of the neonatal screening program.³⁵

Universal Newborn Hearing Screening has now been established in several countries. The reported prevalence of permanent hearing loss identified by newborn hearing screening programmes ranges from 0.3-5/10000.³⁶

The National Health Service Newborn Hearing Screening Program was first introduced in England in 2001. This aimed to identify neonates with a bilateral permanent hearing impairment averaging 40dB or more in the better hearing ear within the first few weeks of life using otoacoustic emissions and automated auditory brainstem reflexes.

Prior to this, the existing universal infant distraction test was being performed at the age of 7-8months. The evidence in the literature at the time had suggested that introducing screening at an earlier age would likely detect any deficits earlier, resulting in earlier intervention, increasing the potential for speech and language acquisition.³⁷

A recent systematic review carried out in Canada researched the evidence for early identification of hearing loss.³⁸ There are areas in Canada that do not as yet have a universal hearing screening program and the average age of detection of hearing loss is 24 months of age, when there is already an established language delay. This is contrasted with an average detection age of 3 months with intervention within 6 months in those children that are screened. Furthermore there is evidence that without intervention, children with hearing loss develop

predictable deficits in speech and language acquisition. The recommendations from this systematic review are that hearing screening should be provided universally for all new-borns that is linked to an integrated system that incorporates diagnosis and intervention.

Neonatal screening has had an impact on the early identification of hearing loss in new-borns, however it is important to note that there are a proportion of children that will go on to develop hearing loss at a later stage in childhood.³⁹ Some groups are sceptical as to the cost effectiveness of universal new born hearing screening for this reason.⁴⁰ This is not necessarily an argument against screening, but highlights the importance of on going vigilance in the detection of progressive childhood hearing loss. Inevitably there are increasing numbers of children with a unilateral hearing loss that being identified. As mentioned previously, not all of these children will go on to develop difficulties with speech and language acquisition, however some of them will. The advent of BAHD processors on a headband has helped to delay surgical intervention in this group until the skull has matured.

2. Skull growth

The growth of the flat bones of the skull is at its most rapid in the first few years of life, and usually reaches its full capacity by the age of 7.⁴¹ In the paediatric immature skull, the bone is often very thin and can be less than 2mm in thickness. The bone is often softer with a higher water content than adult bone and marrow spaces are scattered within the temporal bone.⁴²

These factors are believed to contribute to the longer osseointegration time in children.^{43,44} Surgical techniques need to be modified appropriately to account for this very thin skull and the choice of fixture length has been a subject for debate.²¹ Techniques using bone dust and Gore-Tex® have been reported.⁴²

For those children with a congenital abnormality of the pinna, consideration of later autologous reconstruction should be borne in mind, as this will affect the choice of position for the BAHD.

3. Pre surgical management

The advent of the Baha Softband® has reduced the clinical necessity to implant at such a young age. Christensen et al found that the functional gain in 10 children with a congenital bilateral conductive hearing loss with the Baha Softband® was superior to a traditional bone conduction aid.⁴⁵ This has influenced the management of the very young children with hearing loss in Birmingham and we

now choose to fit children under the age of 3 with the BAHD headband until they are older and more suitable for surgery.

The decision to implant children between the ages of 3 and 5 is taken in a multidisciplinary fashion on a case-by-case basis. The length of the BAHD headband assessment is traditionally longer in children. In our institution, each child will be fitted with a processor on a headband for a minimum of three months before any decision for surgery is made. After this three-month period we rely on reports from the child (if possible) the carers, the teachers and the visiting teachers of the deaf. They are also tested audilogically with and without the band. It is very important that these children use their BAHD in educational settings as well as the home. However we have found that in children with severe learning difficulties, it may not be possible for all children to use the headband for this period of time, particularly those children with severe learning difficulties.³⁰

When children are fitted with a sound processor on a headband this is typically placed in a unilateral fashion even in those children with a bilateral hearing loss. The benefits of binaural hearing have long been discussed in the literature⁴⁶⁻⁴⁸ and for those children with a bilateral hearing loss, bilateral BAHD implantation is offered. Yet it is very interesting that in many BAHD centres these children wear only a unilateral processor on a headband prior to their surgery. We have demonstrated that these children confer additional benefit and quality of life from a second sound processor on their headband.⁵⁰

4. Surgical Challenges

Paediatric BAHD surgery poses various surgical challenges, including timing of surgery, single or two stage surgery, bone thickness and problems encountered with medical co-morbidities in complex children.

Currently there is no clinically accepted consensus for a minimum age of surgery. Initially children were implanted once it was thought that the skull was of a suitable thickness, but there is good evidence in the literature that in the very young, the fixture failure and complication rate is higher.^{21,49} The introduction of headbands has largely removed the necessity of early surgery.

Some centres e.g. Nijmegen use imaging to help determine skull thickness and ideal implant site, however in the United Kingdom this is not accepted practice. There is an unacceptable radiation dose and many children require general anaesthesia or sedation. Another difficulty with the use of imaging is marking the optimal radiological site. Perhaps Cone beam might be of more use in the future as it does have a reduced radiation dose.

Our current practice in Birmingham is to determine the best site for implant position based on the clinical findings at the time of surgery. If thin bone is encountered, various techniques are utilised. Bone dust pate, Gore-Tex® membranes, and longer osseointegration periods. Gore-Tex® is no longer routinely used in Birmingham in children with thin calvarial bone.

Results with the wider implant systems from Cochlea and Oticon show increased fixture stability even in very thin bone.⁵¹

Single stage surgery is standard in the adult population, however in children traditionally BAHD surgery has been performed as a two-stage procedure in order to promote osseointegration. More recently single stage surgery in children has been reported even in the very young.⁵²⁻⁵⁴ Some centres are utilising resonance frequency analysis (RFA) to determine the level of implant stability (and hence osseointegration status). This is commonly used in the field of dental implants and measures the frequency of implant oscillation inside the bone.⁵⁵ A recent study has shown that the BI300 implant system has greater overall stability when compared with previous implants however in order to verify faster osseointegration a larger study would be required.⁵⁶

Lastly, when dealing with a paediatric population, many of the children will have medical co-morbidities including recognised syndromes, craniofacial anomalies and congenital cardiac problems.⁵⁷ This has implications for BAHD surgery as in children the majority is carried out under a general anaesthetic.

5. Complications

Complication are more frequently reported in the paediatric population for most centres implanting children.^{21,49}

Fixture failure: There is evidence that the complications, especially fixture failure is higher in children under the age of 5 and this experience has been reported in Birmingham.^{49,57,58} With the advent of the BAHA Softband ® in 2002 the need for surgery in the very young is now unnecessary until the child is older and hence the skull more mature. It is our standard practice to implant a sleeper fixture in all children where possible that may be used in the event of a fixture failure.

Peri-abutment soft tissue reactions: These have long been a point of intense debate and concern.⁵⁹ An early classification by Holgers is currently the most widely used method of recording these soft tissue problems.⁶⁰ Recent years have seen different abutment shapes and lengths being advocated and recent studies are now reporting that minimal soft tissue reduction and in fact no soft tissue reduction has the best results at present, although longer term results are awaited.

The most recent development includes the launch of a hydroxyapatite-coated abutment from Cochlear®. Experimental studies have shown that the hydroxyapatite coated abutments promote enhanced dermal adherence.⁶¹

Peri-abutment soft tissue reactions are reported to be more of a problem in children. The various theories including hygiene and/ or hormonal issues and poorer socioeconomic status have been implicated.^{43,62-65} Since the children rely entirely on their carers to care for the BAHD and surrounding skin, it is very important that the families, carers and schoolteachers are fully educated into their responsibilities as part of the pre-surgical pathway.

Trauma:

Trauma to the BAHD abutment site is well documented in the paediatric population and is usually implicated in fixture loss.^{43,49,66} Fortunately, more serious complications following trauma are rare, however intracranial intrusion, intracranial haematomas and intracranial abscesses have been described suggesting that in children prone to falls or in those with behavioural problems that perhaps a processor on a headband may be an appropriate choice.⁶⁷⁻⁷²

6. Audit of outcomes

Another challenge for a paediatric BAHD team is the outcome evaluation for children, especially with regards to their quality of life. Evaluating the benefit of BAHDs in children is fraught with difficulty since many children cannot report benefit or complications. Clinicians rely upon the caregiver's perspective when interpreting the impact of the BAHD or headband. In addition to this, the changes in a child's life as they grow and develop also result in large variations in outcome results. The psychological, emotional and educational development is constantly changing so it is not uncommon to have two extremes of results from the same child with a BAHD taken over a period of a few years. These issues clearly introduce a further complexity into the analysis of results for paediatric BAHD centres and clinicians should be mindful of this when interpreting results.

Evaluation of audiological outcomes in this group is equally challenging. The age and cognitive maturity of the child necessitates a variety of audiological testing methods including where necessary: Visually Reinforced Audiometry, Performance Testing or Pure Tone Audiometry. Occasionally it is necessary to obtain Auditory Brainstem Responses under general anaesthesia in the complex child where conventional testing methods are unsuitable. In contrast to adult BAHD practice, the use of speech testing is not routinely performed in children in Birmingham.

Recent developments

Transcutaneous bone conduction aids for use in children

Concern surrounding appearance and peri-abutment skin problems have led to the development of transcutaneous bone conduction devices with similar indications to the skin penetrating or percutaneous devices. They are still relatively new and their results in children remain to be seen.

Transcutaneous passive implant

The Alpha 1 by Sophono™ is a semi implantable system that relies on magnetic coupling between implanted and external magnets.⁷³ This is being trailed in over 100 patients in Germany⁷⁴ and is gaining popularity in the UK.^{75,76} As there is no percutaneous abutment there is less chance of skin reaction and trauma. However the device cannot be used in patients with bone conduction thresholds of greater than 45dB. The experience thus far in Birmingham is that the Alpha 1(M) sound processor is only sufficiently powerful for mild conductive hearing losses at this stage.⁷⁷ Preliminary results from France show good tolerance and patient satisfaction⁷⁸ and a recent study from Nijmegen confirms that the percutaneous BAHD has an output that is 10-15 dB higher than that of the Sophono™ but is effective in children with a unilateral conductive hearing loss.⁷⁹ A further implication is the relative contraindication for an MRI scan. A maximum of 3 Tesla scanner can be used to image the head with a head bandage firmly applied to the area of implantation. If a more detailed scan is required, then the magnet needs to be removed prior to this. In children, the chance of requiring a MRI scan during their lifetime is significant.

Transcutaneous active implant

The latest development has been the Bonebridge system by Med EL⁸⁰ which is an intact skin device with a fully implantable floating mass transducer that is retained within the mastoid temporal bone by two screws that do not rely on osseointegration. An external processor is coupled to internal magnets and the candidacy criteria are similar to that of the Alpha1.⁷⁵ There are no current published trials at the time of writing and the device is not FDA approved for use in children. Should this approval be granted in the future, the considerations for use in children would be the need for a pre operative CT scan to establish sufficient mastoid development and also the above mentioned issues of the implanted magnet and the implications for MRI scanning.

As the complexity of available implants increases so do the considerations for use in children.

Non-surgical bone conduction device

Soundbite is a non-surgical bone conduction device that provides sound via a small piezoelectric device mounted on a removable oral appliance similar to a dental retainer. In patients with unilateral hearing loss, the oral appliance is placed on the hearing side and a microphone is worn in the contralateral external auditory meatus. The sound is then sent from the deaf side to the oral appliance via FM transmission. Studies show that it is well accepted by patients with an acquired single sided deafness and provides improved quality of life.^{81,82} At the time of writing this thesis there are no reports in children, but ingestion or inhalation may be a risk as well as difficulties with fitting due to primary and secondary dentition transition. It may certainly be a more popular option for those older adolescents who refuse any BAHD based on the appearance and self-image.

Quality of life studies in paediatric BAHD

A great deal of research has been done on the evaluation of quality of life after implantation of BAHD in children. The measurement of any improvement in quality of life in the paediatric setting involves a combination of patient and carer responses, depending on the age of the child implanted. The overwhelming evidence in the literature suggests that BAHDS do confer an improved quality of life in children.

There are a variety of questionnaires used:

Infant Toddler Meaningful Auditory Integration Scale (IT-MAIS). A structured interview designed to assess a child's response to sound in everyday environment.⁸³

Glasgow Children's Benefit Inventory (GCBI). A validated questionnaire covering four domains: emotional benefit, physical health, improvements in learning ability and vitality in relation to a health intervention.⁸⁴

Glasgow Hearing Aid Difference Profile (GHADP). A modified version of the validated Glasgow Hearing Aid Benefit Profile questionnaire (GHABP) changed to evaluate existing hearing aid wearers that have a change in hearing aid.⁸⁵

Nijmegen BAHA Quality of Life Instrument (NBQOL)

Visual Analogue Scale of Change in Health Status (VAS). A visual analogue scale used both pre and post operatively to evaluate the change in health status after an intervention.

Daily use of bilateral BAHA Questionnaire (BBQ) questionnaire derived Chung and Stephens⁸⁶ enquiring about daily use of bilateral BAHDs.

Modified Speech Spatial and Qualities of Hearing Questionnaire (SSQ) initially developed by Gatehouse and Noble⁸⁷ and modified for use with children by Galvin.⁸⁸ Evaluates hearing abilities across three domains: speech perception, spatial hearing and other qualities on a VAS.

Children's Home Inventory of Listening Difficulties (CHILD). This is a 15 item questionnaire for the parent or child to fill in regarding communication in typical family communication situations at different distances and in background noise.

Listening Situations questionnaire (LSQ parental). A validated questionnaire used to assess educational and social benefit gained by wearing a hearing aid.⁸⁹ A score of more than +22 would suggest inadequate rehabilitation.

Abbreviated Profile of Hearing Aid Benefit (APHAB). This is a 24 question tool that measures a reduction in hearing disability after the application of a hearing aid. The score ranges from 1 to 99 with the higher score representing greater difficulties.

Table: Summary of quality of life studies in paediatric BAHD

Year	Author	Country	N	Study design	QoL tool	Response rate	Result
2007	Lloyd	UK	71	Retrospective case review of all paediatric BAHD over 12 years	GCBI, GHADP, NBQOL	16.9%	GCBI no worsening of QoL, GHADP reduction in hearing difficulty compared with BC aid and increased time BAHA worn NBQOL more effective, better sound quality
2009	McDermott	UK	115	Retrospective postal questionnaire study of all paediatric BAHD over 15 years	GCBI	73%	GCBI ave +54
2010	Doshi	UK	4	Retrospective case review of BAHD in children with severe behavioural problems	GCBI, VAS	100%	GCBI ave +41.7 VAS ave change in health status +5.6
2012	Dun	The Netherlands	23	Retrospective review of all paediatric BAHD with bilateral CHL over 12 years	Daily use of bilateral BAHA Q GCBI Modified speech spatial and qualities of hearing	91%	GCBI ave +38 Daily use BL BAHA 95% felt BAHA worth the effort 90% used every day Mean SSQ 5.8
2010	Christensen	USA	23	Retrospective case review of all children with BAHD for unilateral SNHL	CHILD	100%	Improvement in mean CHILD score
2010	Ramakrishnan	UK	30	Retrospective case review of all paediatric BAHA over a 9 year period. Small cohort for QOL questionnaires	GBI LSQ	50%	GBI +29 LSQ +17
2011	de Wolf	The Netherlands	38	Retrospective questionnaire study of Paediatric BAHA in unilateral and Bilateral hearing impairment	GCBI APHAB	82%	+ve GCBI in all groups APHAB majority significant clinical benefit

Conclusion

In the UK regular universal neonatal screening programs are detecting hearing loss at an earlier stage. There is increased awareness amongst parents regarding hearing loss and its treatment. The Internet and patient support groups have improved knowledge and highlighted the available options for auditory habilitation. Compared to a decade ago there are many more options for managing hearing loss with a greater choice of both surgical and non-surgical treatments.

We have seen the introduction of the BAHD softband give us not only an indication of potential benefit prior to implantation, but also the ability to influence the age of implantation in children. The BAHD softbands are still a very reliable and accepted form of habilitation until the child has grown sufficiently to endure anaesthesia and to have a thicker more mature skull.

Modernising anaesthetic techniques, simplification of the surgical procedure and centralisation of children's services into specialist centres has made surgery an option for even the most complex children with craniofacial abnormalities and medical co-morbidities.

BAHDs have proved a valuable resource and with the success of expanded indications, the applications are sure to grow. The future technology is very exciting and with the advent of superior processing and implant design, the prospect of further transcutaneous devices, or perhaps even fully implantable devices is achievable during the lifetime of our children.

The Scope of this thesis

The overall objective of this thesis is to assess the special considerations for paediatric BAHD. This is with particular reference to the difficulties encountered in the implantation of young and complex children, both surgical and peri-operative. In addition this thesis looks at the specific quality of life outcome measures in children with a unilateral hearing loss of both a conductive and sensorineural nature.

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Chapter 1-2

Bone-Anchored Hearing Aids versus Conventional Hearing Aids.

R. Banga, R. Lawrence, A. Reid, A-L. Mc Dermott

Abstract

Hearing amplification technology has been evolving since the 19th century. Currently in most audiology departments, the mainstay of hearing rehabilitation is performed with conventional air and bone conduction aids. These are cost-effective, non-invasive hearing aids but are not without their drawbacks. This chapter explores the advantages and disadvantages of conventional hearing aids compared with the bone-anchored hearing aids. Although the bone-anchored hearing aids are a more expensive invasive option, there is increasing evidence that the benefits outweigh the disadvantages. Users report improved quality of life, health status and audiological rehabilitation.

Introduction

Hearing amplification is noted as far back as pre-historic times with animal horns used to amplify speech. At the beginning of the 19th century, early hearing aids in the form of ear trumpets were used to direct sound at hearing-impaired individuals.¹

In 1876, the first bone conduction (BC) hearing aid was described by Prof Paladino at the University of Naples. It consisted of a metal rod; one end was placed on the speaker's larynx and the other end against the teeth or mastoid of the listener. The device was called a Fonifero.² This still required a direct physical link between the speaker and the listener in order to function. See figure 2 in the chapter by Mudry and Tjellström.³

In the 1920s with the advent of electricity and with Alexander Graham Bell's work on the telephone, hearing aid technology had advanced and the electric hearing aid had become commercially available.⁴ In the 1960s, the transistor technology made the 'behind the ear' air conduction (AC) hearing aids a reality¹.

For those patients requiring a bone conductor hearing aid, there was an electromagnetic vibrator designed to press against the mastoid by a tight steel headband. This was uncomfortable and very unsightly. The hearing spectacles were considered a more cosmetic alternative (figure 1).

Modern day conventional hearing aids have three main components: A microphone that converts sound energy to electrical energy, an amplifier and a receiver to convert the modified and amplified electrical impulses back into to sound.

These conventional hearing aids can be separated into two types, those that work by AC and those that work by BC.



Figure 1. The hearing spectacle. http://www.thedeafblog.co.uk/2009/03/hidden_hearings_spectacle_hear.html.

This chapter aims to discuss the merits of both conventional AC and BC hearing aids and compare them with the bone-anchored hearing aid.

Conventional Air Conduction Devices

An AC device uses an ear mould or a 'dome' coupling to deliver amplified sound directly into the ear canal. The receiver and microphone are traditionally worn behind the ear and coupled to the ear mould by a small length of plastic tubing (figure 2). Smaller 'in the ear' AC hearing aids are also available. A bespoke device is made to incorporate the receiver and microphone in a plastic shell which is fitted directly into the conchal bowl or the ear canal, making it almost invisible.

There are also body-worn versions, where the microphone is worn on the clothing and the receiver is attached to the ear mould. The receiver and microphone are attached to each other by a wire (figure 3).

Conventional Bone Conduction Devices

With the BC aids, the receiver is attached to an oscillator that vibrates. BC hearing aids may be either a body-worn or ear level device (incorporating the microphone and amplifier) attached to an oscillator that vibrates in relation to the sounds entering the hearing aid microphone.

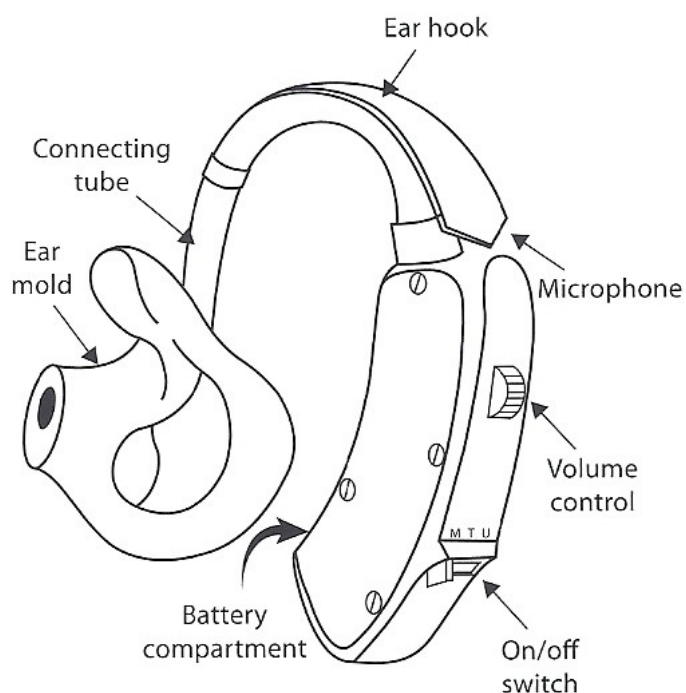


Figure 2. Behind the ear AC hearing aid.
<http://www.bc-childrens.ca/Services/ClinicalDiagnosticFamilyServices/Audiology/Forfamilies/hearingaids.htm>.



Figure 3. Body-worn hearing aid. <http://www.voonradio.com/body.php>.

The oscillator is placed against the skin overlying the mastoid process and is held in place either by a sprung headband, or it is worn on an elastic headband (such as a sports sweatband or the Baba Softband®). In the latter case, the bone

oscillator and the hearing aid are held in place by Velcro pads that attach to the headband.

In the ear level device, the hearing aid is attached to one side of the headband and the oscillator to the other. A wire is threaded over the headband to connect the hearing aid to the oscillator (figure 4). In patients with poorer BC thresholds, a more powerful body-worn device can be used.



Figure 4. BC hearing aid. <http://boingboing.net/2006/01/09/musician-requests-tr.html>.

BC aids are most commonly indicated where an AC aid is not suitable, for example in patients with chronic otitis externa, congenital ear canal atresia or in patients with anatomical abnormalities following mastoid surgery.

Bone-Anchored Hearing Aids

In 1980, Tjellström and his team described the combination of osseointegration and direct BC with a hearing aid.⁵ In 1987, the first semi implantable BC hearing aids became commercially available.⁵⁻⁷ They consisted of a titanium fixture and abutment secured to the skull by osseointegration. The sound processor attached directly to the abutment (figure 5).

The percutaneous bone-anchored hearing aid (Baha[®]) has since proven to be an effective method of rehabilitation for patients with both unilateral and bilateral conductive and mixed hearing loss.^{8,9} A summary of the evidence regarding bone anchored and conventional hearing aids can be found in table 1.

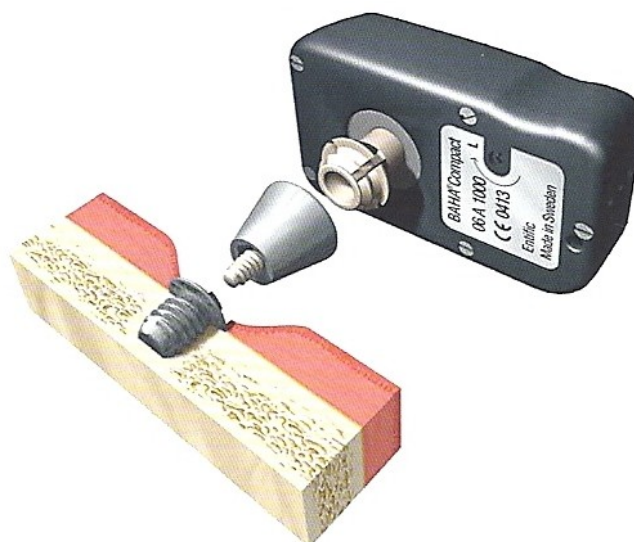


Figure 5. Baha. <http://www.ent.uci.edu/BAHA.htm>.

Advantages of Baha versus Bone Conduction and Air Conduction Hearing Aids

Sound Quality

Compared to the Baha, sound clarity is compromised using a conventional BC device, particularly at the low and high frequencies. This is a result of sound dissipating within the hair, skin and muscle tissue before reaching the bone of the mastoid.

Christensen et al.¹⁰ found a statistically significant audiological improvement in children with a Baha compared with traditional BC aids. Interestingly, they found that there was no significant difference in these children between the Baha Softband and the implanted Baha. Studies from both Nijmegen and Birmingham have also shown that patients report a subjective improvement in sound quality using the Baha compared with traditional BC aids.^{8,11,12}

Better speech recognition is mentioned most frequently as an advantage of Baha over conventional hearing aids, particularly in noisy surroundings.⁸ Two smaller studies reported a statistically significant improvement in questionnaire results with the Baha compared to BC aids. Interestingly, this improvement was not demonstrated in the speech discrimination test on the same patients.^{8,13}

With a Baha, sound bypasses the 'problem areas' - the outer and middle ear, and maximises the level and quality of sound that reaches the cochlea.

Table 1. Summary of the evidence for Baha

Reference	First author	Methods	Results
8	Mylanus	Questionnaire	Improved speech in quiet and noise and improved sound quality with Baha
10	Christensen	Audiometric tests	Improved functional gain over BC aids
11	McDermott	Nijmegen questionnaire	Baha better in all audiological and patient respects
12	Mylanus	Questionnaire and audiometric tests	Small improvement with speech in noise with the Baha but majority of patients preferred Baha
13	Bonding	Audiological audit	Baha superior to BC and AC aids, but speech discrimination in noise was the same with Baha, AC and BC
14	Oeding	Hearing in Noise Test (HINT) and abbreviated profile of hearing aid Benefit questionnaire (APHAB)	Directional microphone was much better in Baha for Speech in Noise Test
16	Hol	Audiometric tests	Better speech in noise
17	Hol	APHAB	Improved speech in noise with Baha as a CROS aid. Significant benefit with the APHAB
18	Lin	Subjective benefit questionnaire, HINT	Benefit for most patients with a degree of SNHL with Baha
19	Hol	Audiological tests, Directional hearing questionnaire, APHAB	Benefit with Baha, BC and AC
20	Bosman	Audiological tests	Directional hearing and speech in noise improved with Baha
21	Hol	Audiological tests	Baha Softband as good as other aids
22	Doshi	Glasgow children's benefit inventory (GCBI)	Improved quality of life with Baha
23	Snik	Audiological tests, questionnaire	Baha was better than previous conventional aids
24	Hol	Aided free field speech in noise and quiet	Improved speech in quiet and speech localisation
27	de Wolf	Glasgow benefit inventory (GBI), APHAB, Nijmegen cochlear implant questionnaire, Hearing handicap inventory for the elderly	Better quality of life
28	Hol	6-item short form health survey, EuroQol-5D, Hearing handicap and disability inventory	Improved quality of life
29	Kunst	GCBI, Listening inventory for education questionnaire	Subjective benefit with Baha
31	Dutt	GBI, Health status	Benefit with bilateral Baha and improved health status
34	McDermott	GCBI	Benefit with Baha

For conditions characterised by fluctuating hearing loss but stable BC thresholds (typically middle ear effusions and infection), the sound perceived from a BC device will be consistent, whereas sound perceived with an AC device will be heavily dependent on AC thresholds at the time. This creates an issue when programming the AC aids, as most fitting formulae such as Desired Sensation Level or National Acoustics Laboratory take the AC and in some cases the BC levels into account when prescribing the amount of gain needed. If these fitting formulae calculations are constantly changing, it is impossible for the AC aid to function optimally. When compared to conventional BC aids, the Baha has been shown statistically to have as much gain as a BC transducer at 500, 1,000, 2,000 and 4,000 Hz in children.¹⁰

Sound Localisation

The ability to localise sound is severely diminished with a BC aid. The microphone may be situated on clothing or may transmit sounds from the contralateral side. This creates unwanted background noise, leading to an unnatural listening condition. Ear level Baha sound processors have the microphone on the same side, which is a more natural listening condition.

Directional microphones in conventional AC hearing aids can provide a greater signal-to-noise ratio than omnidirectional microphones to improve speech recognition in the presence of background noise. The Baha now has the advantage of directional microphones. These have been shown to provide a statistically significant improvement in speech in noise recognition.¹⁴

The management of unilateral hearing loss has been controversial. Recent studies however have shown the efficacy of the Baha as a contralateral routing of signal (CROS) device by eliminating the head shadow effect.¹⁵⁻¹⁷ In fact, Baha has been shown to provide better amplification in subjects with unilateral hearing loss than the conventional CROS aid.^{18,19}

For patients with bilateral hearing loss, results have shown that directional hearing and speech recognition in noise improves significantly with a second Baha, resulting in binaural hearing.²⁰

Comfort

Comfort is an issue, particularly when the BC aid is worn on a sprung headband, since this delivers a degree of pressure on the skull, which can be uncomfortable if worn for long periods of time. A major development in BC was the introduction of the Baha Softband. The Baha attached to an elasticised headband has proven to be a very successful, attractive and acceptable method of hearing rehabilitation for

younger children.²¹ Although there are still pressure and comfort issues with the Baha Softband, the position can be easily altered to a different site on the skull.

A significant advantage of the Baha is its direct attachment to the bone. This renders the device 'weightless' from the patient's perspective. There is no pressure applied to the skin or soft tissues. Our early experience from the use of Baha in children, particularly those with severe disability, suggests that this perception of 'weightlessness' may be responsible for increased use of the Baha in this subset of children.²²

Consistency is a particularly troublesome problem when using a BC hearing aid in young children. It is very difficult to prevent the bone vibrator from slipping from the mastoid process. This problem is not encountered with the Baha system.

Cosmesis

Although some BC aids can be disguised to look like hair bands or sports sweat bands, the overall appearance of the Baha is far more cosmetically acceptable than that of a BC aid. It is our experience that teenage children, particularly boys, have a reluctance to wear any BC aid due to concerns regarding their self-image. These children will have the same reluctance to wear a Baha Softband/headband, but they are more accepting of the percutaneous Baha. In studies comparing the Baha with conventional aids, visibility has been a negative factor, but still considered more favourable than conventional aids.^{8,11}

Practical Fitting Issues

There are practical fitting issues that disadvantage AC aids. They require an ear mould or domed coupling to be placed in the ear canal. If there is discharge present in the ear, this will inevitably block the ear mould and lead to a failure of the amplified sound to reach the ear canal. Furthermore, the presence of an infection is a contraindication for impressions to be taken of the ear for the manufacture of ear moulds.

The presence of the ear mould or domed coupling in the ear can reduce the airflow in the ear canal resulting in a moist ear canal, increasing the risk of ear infection. This in turn can lead to inconsistent use of the device. Studies have shown that patients report a decrease in the frequency of ear infections when using a Baha as compared to conventional AC hearing aids.^{11,12,23,24}

Children require multiple mould taking as they grow to ensure that the mould is tightly fitting for best use. Cooperation in obtaining an accurate impression for the hearing aid mould can be challenging, and complications have been reported.^{25,26}

Quality of Life

Subjective benefit from patients with Baha has repeatedly been shown to be improved when compared to their prior conventional aids. This benefit has been shown in children, adults, patients with learning difficulties and more recently older users.^{22,27-30} A subjective improvement in a general state of health (and hence quality of life) has been reported with both unilateral and bilateral Baha use.³¹ For those patients asked to state a preference for hearing aid, the overwhelming majority prefer Baha.¹¹

Disadvantages of Bone-Anchored Hearing Aids*Surgery*

This is an invasive procedure and carries with it both surgical and anaesthetic risks. Surgical implantation of a fixture and percutaneous abutment are necessary for attachment of the Baha sound processor. In the adult population, it is possible to perform the surgery under a local anaesthetic technique as a 'day case procedure', thus reducing the risk to the patient.

In the paediatric population, it is more common for surgery to be performed under general anaesthesia, and often two procedures are advocated 3 months apart to allow for osseointegration. In addition to this the paediatric population is a more complex group of patients with complex anaesthetic needs. A large proportion of children will have recognised syndromes, cardiac problems and other medical comorbidities, making them a challenging group to anaesthetise.³²

Osseointegration Time

The sound processor is commonly fitted after a minimum of 6 weeks following surgery in adults, and invariably 3 months is allowed to elapse between first- and second-stage surgery in children.³³ These timings are intended to allow good osseointegration of the fixture and the bone. This may be longer if surgical difficulties are encountered, most commonly thin calvarial bone at the time of fixture placement.³⁴ There is much research into the strength of osseointegration, and it is likely that these time delays to fitting the aid will reduce.³⁵

Soft Tissue Complications

The maintenance of the soft tissues surrounding the Baha abutment site is vitally important to prevent inflammation, infection and ultimately fixture loss.^{9,34,36-38} In

younger Baha wearers and some adults, a second person is required to ensure that the abutment site is kept clean and healthy.

Trauma

A significant number of patients, particularly children, sustain trauma to their Baha. Fortunately, the sound processor is the most common component to be damaged. Serious trauma to the abutment site is very rare, but has been reported in the literature.^{39,40}

Conclusion

Conventional hearing aids provide the mainstay of hearing rehabilitation and are usually the first line management of hearing loss. They are cost-effective and readily available to most audiology departments. The Baha is a more expensive alternative that requires an invasive procedure(s) and has potential for postoperative complications. Despite these issues, there is increasing evidence that the advantages of Baha outweigh these recognised problems, and users both adult and children report improved quality of life, health status and audiological rehabilitation.

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Chapter 1-3

Bone Conduction and Bone Conduction Hearing Aids.

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Abstract

The phenomenon of bone conduction has become better understood during the past century and over the past three decades numerous clinical and audiological studies have been described in relation to various bone conduction hearing aids. The understanding of osseointegration has led to further developments and the introduction of bone anchored hearing aids. The aims of this article are to provide the reader with an understanding of bone conduction and the evolution of the bone conduction hearing aids, with particular reference to bone anchored hearing aid implant systems.

Physiology of bone conduction

During the past century bone conduction and its physiology has become more fully understood.¹

Von Békésy discovered that the mode of cochlear hair cell excitation was identical irrespective of whether the sound wave pathway was via air or bone conduction.² Later in 1966, Tonndorf hypothesised that bone conduction was the result of contributions from three mechanisms: bone compression, inertia bone conduction and the sound energy of the external ear.

The concept of Bone compression was postulated by Reitjö in 1914.³ He suggested that compression of the inner ear contributed to sound perception by bone conduction. This was later supported by the experiments carried out by Herzog and Krainz in 1926.^{4,5} The bone compression theory was based on the knowledge that vibratory energy reached the cochlea and caused both compression and expansion of the cochlear bony structures. Since the fluid within the cochlea is not compressible, the forces result in fluid displacement. The scalae have different dimensions: the scala vestibuli being larger than the scala tympani. Bone compression results in greater displacement of fluid in the larger scala vestibuli. This displacement of fluid is further emphasised by the semicircular canals and vestibule. This results in downwards displacement of the basilar membrane.⁶

Finally, it is thought that the compliance ratio between the round window and the oval window is 20:1, thus compression results in comparatively more movement at the round window. Movement of the basilar membrane is thus further amplified.

Since basilar movement is greatest at the basal turn, sound perception by bone compression contributes mostly to the high frequencies.^{7,8}

The second mechanism of inertia bone conduction is based on the concept that the incus and malleus vibrate as a single unit. This unit in turn moves as a pendulum within the skull which is also vibrating. The resultant motions between the ossicular unit and the skull displace the stapes, stimulating the cochlea. This is similar to the air conduction pathway.⁹

Lastly, sound energy from the vibrating skull radiates to the bony and cartilaginous walls of the external auditory meatus. This acoustic signal is processed via the air conduction pathway.^{10,11} (Figure 1)

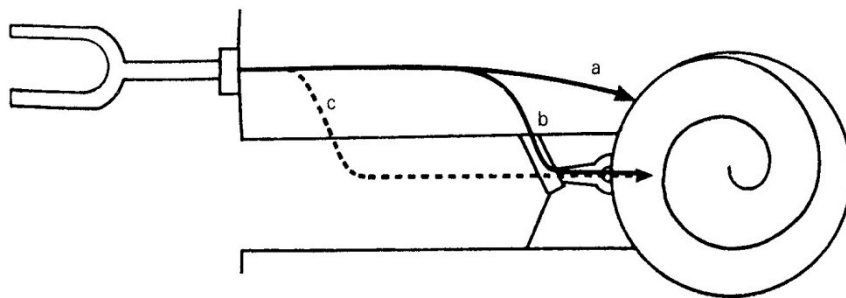


Figure 1. The mechanisms of bone conduction¹¹

More recently the role of the cerebro-spinal fluid (CSF) has been recognised. Any vibrations in the CSF resulting from skull vibration can reach the cochlea directly and in turn induce additional longitudinal fluid waves providing the oval and round windows are mobile.¹⁰

History of bone conduction hearing aids. (BC hearing aids)

Hearing amplification is noted as far back as pre historic times with animal horns used to amplify speech. At the beginning of the 19th century, early hearing aids in the form of ear trumpets were used to direct sound at hearing impaired individuals.¹² In 1876 the first bone conduction hearing aid was described by Professor Paladino at the University of Naples. It consisted of a metal rod; one end

was placed on the speaker's larynx and the other end against the teeth or mastoid of the listener. The device was called a Fonifero.¹³

In the 1920's with the advent of electricity and with Alexander Graham Bell's work on the telephone, hearing aid technology had advanced and the electric hearing aid had become commercially available.¹⁴ Traditional BC devices were often adapted from existing Body Worn (BW) type air-conduction hearing aids that consisted of an ear level receiver linked by a wire to a body worn amplifier. Typically the receivers of these aids would simply be replaced with a vibrating receiver that would be mounted on a tight fitting headband to hold the bone vibrator onto the mastoid.

The relatively large distance between the microphone and receiver was beneficial in that higher amounts of gain could be applied with less chance of feedback but the cumbersome nature of a BW device was considered cosmetically unacceptable.

In the 1960's the transistor technology made the 'behind the ear' air conduction hearing aids a reality and following the widespread introduction of ear level air conduction hearing aids came the introduction of ear level BC aids and spectacle mounted BC aids. The hearing spectacles were considered a more cosmetic alternative (Figure 2).¹⁵



Figure 2. The hearing spectacle¹⁵

Since 1987, the bone anchored hearing aid device (BAHA) has become commercially available.^{16,17} Bone anchored hearing aids are now an accepted form of auditory rehabilitation for adults and children. As indications and surgical techniques have evolved, so has the teamwork between surgeons and audiologists. In the United Kingdom, centres undertaking BAHA work usually have well defined Multidisciplinary teams.¹⁸

Conventional bone conduction hearing aids (BC hearing aids)

Bone-Conduction (BC) hearing aids are typically used for patients suffering with conductive type hearing loss and otological conditions that prevent them using an air-conduction (AC) type hearing aid. Common conditions observed in users of BC hearing aids include chronic otitis externa, chronic suppurative otitis media, otosclerosis and congenital/ acquired abnormalities such as atresia and microtia. Current day BC hearing aids have progressed very little in design since the 1960's and to date remain in the form of either the spectacle BC aids or a body worn/ear level processor. The sound processor, incorporating both the microphone and amplifier is attached to an oscillator that vibrates in relation to the sounds entering the hearing aid microphone and subsequently transfers the sound through the bone to the inner ear. The efficiency of this sound transference will depend on a number of factors including thickness of the skin/soft tissue and the density of the bone through which the signal travels to the cochlea. Inevitably sound will be lost as it is dissipated through the skin and soft tissue and back through the headband holding the vibrator in place. Previous studies have suggested transcranial attenuation may be predicted to be 5-10dB, however this can vary between 0-30dB at any frequency.¹⁹

In recent years BC aids have become more common in the treatment of patients with unilateral sensorineural deafness, often described as single sided deafness (SSD). In such cases the BC device utilises transcranial crossover as a mechanism for transferring sound from the poor ear via bone conduction to the side with a normal functioning cochlea.

Despite the lack of progression in appearance, the technology in sound processing has improved significantly over the last 10 years.²⁰ The United Kingdom (UK) introduction of the Widex Senso digital hearing aid in the mid 1990's paved the way to the introduction of digital hearing aids and a new era in hearing aid technology. Early production digital aids demonstrated significant progress in technology over analogue predecessors offering wide dynamic range compression and non-linear gain alongside digital signal processing and digital feedback suppression. To compliment this technology, the use of digitally programmable devices with multiple channels and bands became widespread within the market enabling the Healthcare professional to actively adjust the hearing aids to meet validated prescription formulae and the subjective needs of the patient. This technology is now available across a range of BC products.

The Bruckhoff Hannover range of spectacle mounted and headband mounted BC devices are well recognised in the UK and offer a modern day solution for patients requiring a conventional BC system.²¹ Alternative systems include the Coselgi spectacle BC devices²² and the recent introduction of the BHM-Tech Contact-Mini²³ targeted specifically at the Paediatric market.

Unfortunately BC hearing aids remain unpopular for a number of reasons: These aids generally have a lack of aesthetic appeal for the patient. Although they can be disguised to resemble sweatbands or 'Alice band'-type hair bands they remain visible and often children, particularly boys, are reluctant to wear any BC aid due to concerns regarding their self-image. The 'sprung' headband causes pressure on the skull and soft tissues and causes discomfort. Often users will try to alleviate this by removing the aid for short periods of time during the day. It is difficult to achieve consistency in using the BC aid as it will often slip off the mastoid process and this results in an intermittency of the sound.

Despite these issues the BC aid is still favoured by some patients who suffer with a temporary or fluctuating larger conductive hearing loss and those who opt not to have an implanted aid.

New developments in BC aids: The TransEar®

A newer alternative to the traditional BC device is the TransEar®²⁴ developed by the Ear Technology Corporation.



Figure 3. The TransEar®²⁴

The TransEar® has the appearance of a conventional behind the ear (BTE) hearing aid with a custom shell made to fit in the ear canal in place of the

conventional ear mould. The custom shell houses a miniature bone conduction oscillator that is positioned in the shell to make contact with the bony portion of the ear canal. It uses a unique high frequency bone vibrator which has a peak output frequency of 2100-2300Hz, in contrast to conventional BC aids that peak near 800Hz. As with a conventional BC aid, sound is picked up from the microphone of the BTE device and then transferred into mechanical energy that drives the oscillator which in turn transfers sound vibration through the bony part of the ear canal directly to the cochlea²⁴

This innovation in BC technology offers an alternative non-surgical solution to the conventional BC aid that is both cosmetically more appealing and has the potential to provide better access to high frequency sound, however as yet there are few published clinical trials with this device.

Bone anchored hearing aids

The major step in bone conduction hearing aid development was the direct transmission of sound to the bone via a rigid implant. The first bone-anchored hearing aid was implanted in 1977 by Anders Tjellström.²⁵ In 1987, the first semi-implantable bone conduction hearing aids became commercially available.^{16,25-27} It consisted of a titanium fixture and abutment secured to the skull by osseointegration. The sound processor attached directly to the abutment. The introduction of the BAHA was not only a huge step forward in terms of comfort and cosmetics but also provided significant improvement in transference of sound and has since proven to be an effective method of rehabilitation for patients with both unilateral and bilateral conductive and mixed hearing loss.^{28,29}

Despite this progress, the technology in these devices has until recent years often fallen significantly behind the technology available in air conduction hearing instruments.

There are currently two percutaneous bone anchored hearing aid systems available in the United Kingdom; Cochlear™ Baha® implant system and the Oticon Medical Ponto implant system providing a range of processors to meet the audiological needs of patients with bone conduction thresholds ranging from -10-65dBHL.³⁰ These modern devices are programmed using manufacturers fitting software that facilitates the use of technology found in the most advanced air conduction hearing aids.

Benefits of fitting software

The fitting software used for the new generation of BAHA offers prescription-based fitting which provides an excellent base for initial programming and fine-tuning. This gives a much more individualised, accurate fitting which is tailored to the patients hearing loss through the multiple channels which can be individually modified to ensure the appropriate level of gain is applied across each frequency to meet the individual patients needs. This should lead to quicker acceptance of the new aid along with improved hearing aid performance.

In addition the software enables the selection of a number of options or features of the aid that are considered beneficial in improving the listening experience for the hearing aid user. An overview of these features is provided below:

BC direct

BC direct is a new technology developed by Cochlear™ to provide a more accurate measurement of bone conduction thresholds and improve Baha® fitting outcomes. BC direct measures the softest sounds the patient can hear directly through the BP100 sound processor (similar to the conventional measurement of bone conduction during an audiometric test) and is measuring the effectiveness of the direct transmission of sound from the sound processor to the cochlea. A limitation when measuring bone conduction with an audiometer is that the sound must pass through the skin and soft tissues before reaching the bone and then the cochlea. BC direct takes into consideration the skin thickness, distance of the implant from the cochlea and transcranial attenuation. It provides an improved method of compensating for the variability in individual patients with a Cochlear™ Baha®.³¹

Directional Microphones and Automatic Adaptive Directionality

Directional microphones have been used successfully in hearing aids for over 30 years. Early studies demonstrated significant benefits in signal to noise ratio of 3-4dB advantage in hard of hearing listeners using directional microphones compared with those using omni-directional microphones.³²

Similar benefits have been observed by in BAHA users where significant directional advantages were observed between subjects with unilateral sensorineural hearing loss when comparing responses using directional and omni-directional microphones of the Cochlear™ Baha® Divino.^{33,34}

The introduction of digital processing in BAHA processors provided the opportunity to introduce more complex directional systems aligned to those currently found in modern air conduction hearing aids.

Both the Cochlear™ Baha® solution and Oticon Medical manufacturers currently supply products offering automated and adaptive directionality whereby the sound processor will analyse the incoming signal and automatically switch between Omni and directional microphone modes based on whether the signal is interpreted as noise or speech, then where interpreted as noise, the aids adapt the polar plot of the microphone continuously to provide optimum noise reduction regardless of the changing position of the noise source.^{35,36}

Automatic Noise reduction

Digital signal processing enables the analysis of the incoming signal and the opportunity to reduce unwanted noise and improve acceptability of the amplified signal to the listener. This is achieved using an algorithm that interprets a signal that is modulated and varies in intensity over time to be primarily speech and a signal that is steady and unvaried to be primarily noise.

When detecting steady state noise the gain of the device will be automatically reduced. Therefore the introduction of noise reduction does not increase the signal to noise ratio because the gain for all sounds is decreased.^{35,37} The benefit of this technology is therefore not observed in speech intelligibility but has been reported to provide improved sound quality³⁷ and observed benefits in comfort in noisy listening situations.³⁸

Data Logging

The concept of data logging enables the clinician to monitor and observe recorded data relating to individual's usage of the device. This information can provide the clinician with an overview of the settings or programs commonly selected, the volume control level and the overall hours of use. This information is often useful during the period of adaptation to the device and can be particularly useful in the counselling process. It provides the clinician with a true reflection of hearing aid usage and as such will often provide an accurate insight into the root cause of problems reported in the early stages by the user. In BAHA users this information can prove invaluable, particularly in children and adolescents where aesthetic issues often result in the removal of the device and a reluctance to wear it.¹⁸

Data learning

Data learning takes the concept of data logging a step further whereby based on user preferences the hearing aid will ultimately learn these preferences and select them as the default start up settings. With use of a BAHA this concept is at present only applied to volume control learning. Whilst such features can be beneficial, it is

essential that consideration is given to the appropriate timing of introducing such features to a device. It is commonly accepted that new users of any hearing aid device will find the reintroduction to sound somewhat alien to them and as such there will be a period of adaptation. New users will typically turn their hearing aids down during the initial period of adaptation but as time progresses they will increase the volume to a level one would expect to be close to prescription. There is therefore a risk that if data learning was introduced too early, the settings of the device would be adjusted before the user had the appropriate time to adapt.

Frequency Transposition/Frequency Compression

The role of a hearing aid has traditionally been to compensate for the users hearing loss by providing a prescribed amount of gain to overcome the hearing deficit. The concept of this is reasonable and has proved successful in users who have non-complex hearing loss.

In considering a patient with a flat 70dBHL hearing loss across frequencies 250Hz-6kHz it is easy to visualise that by applying a simple prescription you can apply equal gain across each of these frequencies to compensate for that loss.

Equally with a mild sloping hearing loss progressing from 30dBHL to 70dBHL across the frequencies 250Hz-6kHz using multichannel programmable hearing aids it is equally easy to visualise that by applying a simple prescription you can apply varying gain across each of the frequency bands to again compensate for the hearing loss.

Unfortunately when transferred to patients with complex hearing loss this concept is flawed and often leads to unsatisfactory end user experience.

A major challenge is experienced when considering users with severe high frequency hearing loss. Users presenting with severely sloping hearing loss typically demonstrate normal hearing thresholds between frequencies 250Hz-1kHz followed by a rapid deterioration from 2kHz-6kHz. In such cases it is not uncommon for thresholds above 3KHz to be in excess of 100dBHL or in extreme cases unobtainable and hence amplification at such frequencies will provide minimal benefit and may prove to be detrimental due to the effects of off-frequency basilar membrane stimulation.

The main impact of this problem is difficulty in recognizing certain speech sounds, such as the fricative consonants /f/, /s/, and /sh/ and difficulty identifying high-pitched sounds, such as birdsongs, alarms, and some musical sounds. One potential solution to this problem is the concept of frequency shifting often described as frequency transposition. The mechanics of this can be simplified and explained as a shifting of a range of high frequencies that were previously

inaudible to the user to an area of lower frequency that is audible to the user. Whilst this method has proved beneficial in providing the user with access to sounds at frequencies previously inaudible the transposed range of frequencies will overlap the lower frequencies present in the input signal potentially resulting in sound distortion.

To avoid such overlap the concept of frequency compression can be applied. This technology has been used successfully by Phonak™ and was introduced as Sound recover in their Naida® and Nathos® range of air-conduction hearing aids. Sound recover allows a selected range of frequencies to be progressively compressed into a narrower range. Consequently sound will be compressed to an adjacent area where there is less cochlear damage and hence where it will become audible to the listener without overlapping other frequencies and producing further distortion.

As yet this technology is not widely available in existing BAHA devices, however given the limited high frequency response of any BC device one would expect this to be the obvious advance in technology for manufacturers of BAHA and other BC devices. This is very soon to be addressed by BAHA manufacturers.

Active Feedback Cancellation

Probably the most common complaint of hearing aid users over the years has been acoustic or mechanical feedback, often described by the user as ‘whistling’. Modern BAHA systems will combat this using active feedback cancellation. This technology has been commonly used in air conduction devices and works on the following principle:

When feedback occurs, the device will detect it, and subsequently the device will introduce an identical tone that is 180 degrees out of phase with the feedback tone thus resulting in phase cancellation. The benefit of this over alternative methods of feedback suppression is feedback is reduced without the need to reduce hearing aid gain. As a result this technology enables the provision of additional gain which should provide the patient with increased audibility compared to previous generations of BAHA sound processors.³⁹

BAHA Clinical Outcomes

Studies from both Nijmegen and Birmingham have shown that patients report a subjective improvement in sound quality when using the BAHA compared with traditional BC aids.^{5,6,28,40}

A retrospective study using the Nijmegen questionnaire found an overwhelming 83% preferred their BAHA to their previous hearing aids, 78% of users liked the

quality of sound from the BAHA.⁴⁰ A statistically significant audiological improvement was found in children with a BAHA compared with traditional BC aids by Christensen et al.⁴¹ They also found there was no significant difference in those using the BAHA softband and those with an implanted BAHA.

Patient subjective outcomes have been well reported by many centres. Excellent overall satisfaction with the BAHA when evaluating using the quality of life questionnaire has been frequently described.^{28,42-46}

For patients with a bilateral hearing loss, directional hearing and speech recognition in noise improves significantly with a second BAHA, resulting in binaural hearing.⁴³ Also, the use of bilateral BAHAs significantly enhanced the patient's subjective general well-being and health status.⁴¹

Advantages of BAHA

The BAHA is 'weightless' as it is directly attached to the bone and therefore does not cause discomfort as with conventional BC aid use. This perception of 'weightlessness' may be a reason for the increased use of a BAHA in the cohort of children with severe disability, who refuse to wear a BC aid for anything more than a few moments.⁴⁷

Visibility of the BAHA can sometimes be a cause of reluctance to have it implanted, but it is still considered to be more favourable than conventional aids.^{28,44} It is overall a much smaller device when compared to BC aids and has greater cosmetic appeal.

A BAHA gives a reliable transmission of sound through direct bone conduction, as no sound is 'lost' passing through the hair and soft tissues.

The Xpress unit from Oticon facilitates the sound processor to be both fitted and removed from the abutment without the 'snap' action. This reduces any lateral stresses placed on the abutment and allows the hearing aid to be used by the patient earlier than traditionally practiced. This is a helpful tool for parents and patients that do not feel confident connecting and removing the processor due to the force required. Recent research from Birmingham has shown that fitting of the BAHA earlier using the Oticon Xpress unit did not result in any complications.⁴⁸

Disadvantages of a BAHA

Surgical procedure and considerations

Having a bone anchored hearing aid system is unquestionably an invasive procedure and both surgical and anaesthetic risks should be included in any discussion with patients considering this procedure. It is prudent to trial a bone conductor headband prior to embarking on any surgery. Patients with benefit from

a headband trial can anticipate good audiological outcomes with a BAHA. BAHA surgery in the adult population is less fraught with difficulties and it is commonly possible to perform the surgery in a single stage under a local anaesthetic technique as a 'day case procedure'.

The paediatric population has a large proportion of children with recognised syndromes and additional medical needs making them a challenging group to anaesthetize.⁴⁹ It is usual for surgery to be performed under general anaesthesia in two stages incorporating a three month osseointegration period. This reduces the risk to the patient. Single stage surgery may be considered in children over the age of 10 years⁵⁰ and a few centres advocate single stage surgery in all patients irrespective of age and report good results.^{49,51}

Soft tissue maintenance

The maintenance of the soft tissues surrounding the BAHA abutment site is vitally important to prevent inflammation, infection and ultimately fixture loss.^{28,52,53} In younger BAHA wearers and some adults a second person is required to ensure that the abutment site is kept clean and healthy. Increased soft tissue complication rates are described in paediatric BAHA practice when compared to the adult population.^{50,52,54,55} Soft tissue problems are far more frequently encountered than fixture failure. Significant hypertrophy of the skin surrounding BAHA abutments requires intensive medical treatment and in some cases even surgical skin reduction. Increasing use of the long abutment has been shown to be beneficial in those patients with soft tissue difficulties.⁵⁶⁻⁵⁸

Studies have shown that the majority of BAHA users did not perceive care of the implant site as a burden.⁴⁴

Osseointegration period

The sound processor is commonly fitted after a minimum of 6 weeks following surgery in adults and typically three months is allowed to elapse between first and second stage surgery in children.⁵⁹ These timings are intended to allow good osseointegration of the fixture and the bone. This may be longer if surgical difficulties encountered, most commonly thin calvarial bone at the time of fixture placement.^{54,55,60}

There is much research on the ideal period of osseointegration and it is likely that current time delays prior to fitting the aid will reduce.⁶¹⁻⁶² The introduction of a newly designed implant Cochlear™ Baha® B1300 implant has provided higher implant stability demonstrated using resonance frequency analysis (RFA) than

with previous generation implants.⁶²⁻⁶³ There is evidence supporting the fitting of BAHA sound processors as early as four weeks post implantation without complication.^{64,65}

BAHA is not as widely available as conventional hearing aids and hence may require travel to a centre for the assessment, surgery and follow-up appointments.

Trauma

A significant number of patients, particularly children, sustain trauma to their BAHA. Fortunately, the sound processor is the most common component to be damaged. Serious trauma to the abutment site is very rare, but has been reported in the literature.^{66,67}

Practical considerations

Imaging causes concern for patients and radiologists when a Baha is present. The traditional percutaneous titanium abutments are not contraindicated for magnetic resonance imaging (MRI). It is only the sound processor that needs to be removed. The abutment free Sophono™ has magnets within a titanium casing. This system is not compatible with MRI. All the BAHA implant systems cause artefact on computerised tomography (CT).

New Developments in Bone Conduction Implants

Sophono™

2010 saw the renewed introduction of the United Kingdom's first abutment free bone anchored hearing aid: Sophono™. This exciting addition to the choice of bone anchored hearing aid products involves the insertion of an implant consisting of two magnets, hermetically sealed in a titanium case and fixed by titanium screws. This implant is completely passive and ideally should not need to be replaced.^{15,68}

With the new abutment free Sophono™ implant system, only one surgical procedure is necessary. Furthermore, no subcutaneous skin reduction should be required and it is unnecessary to remove the hair follicles as with traditional percutaneous BAHA surgery. This has cosmetic implications particularly for the male population with shorter hair. The abutment free Sophono™ does not rely upon osseointegration. Once the surgical site has healed, the sound processor is ready to be fitted.

Soundbite™ Hearing System

Sonitus Medical™ has developed the SoundBite™ Hearing System. It is marketed as the world's first non-surgical and removable bone conduction hearing system designed to transmit sound via the teeth of the upper jaw. The SoundBite™ Hearing System consists of both a BTE unit which houses the receiver, a wireless transmitter, and attached microphone that will capture the sound and transmit a signal wirelessly to a removable In-The-Mouth (ITM) hearing device that will transfer the signal by sound vibration conducted through the teeth and the bone of the skull to the cochlea. The SoundBite™ Hearing System is intended for patients who suffer from single sided deafness, conductive hearing loss, or mixed hearing loss and seek a non-surgical, non-invasive, hearing solution.

Early studies (performed by Sonitus Medical™) indicate high levels of satisfaction in excess of 95% and no evidence of long-term medical or dental effects as a result of using the device. In January 2011 the product was given food and drug administration (FDA) approval in the United States for use in the treatment of single sided deafness.^{69,70}

Conclusions

Bone anchored hearing aid outcomes both audilogically and clinically have proved to be successful and indications for such hearing devices have continued to increase. Advances in hearing aid technology have resulted in major progression in the function of bone conduction hearing aids. As technology improves, not only will the processing capabilities and listening experience for the end user improve, but devices are expected to become significantly smaller and cosmetically more acceptable.

Testing of the early bone anchored hearing aids was performed on the teeth of the upper jaw yet it is only recently gained popularity. The more recent and exciting developments of the abutment free implants and the in the mouth' hearing systems have become not only a reality, but a viable option for patients with hearing loss. The integration of both Wi-Fi and Bluetooth technologies have also contributed greatly to the modernisation of such devices.

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Chapter 2

Bone Anchored Hearing Aids in very young children.

R. Banga, A. Reid, D. Proops, A-L. McDermott

Abstract

Objective: To evaluate the morbidity of bone anchored hearing device surgery in children below the age of 5 years.

Study design: Retrospective case analysis.

Setting: Birmingham Children's Hospital, UK (tertiary referral centre).

Patients: All children below the age of 5 years who were fitted with a bone anchored hearing device in the last 18 years.

Main outcome measures: Medical co-morbidity, fixture failures, significant skin reactions

Results: 53 patients aged less than 5 years at the time of implantation were identified. The age range was 2 years to 4 years 11 months.

60% of the children had a significant medical co-morbidity and the overall fixture failure rate in this young cohort was 33.8%.

Significant skin reactions requiring revision surgery were encountered in 32% cases.

Conclusions: The use of the bone anchored hearing devices in children under 5 has previously been controversial. In the Birmingham series, there was an increase in morbidity in this young patient group and the fixture failure rate was high. Over the past 9 years, the Baha® Softband™ has been the treatment of choice for these very young children and surgery is now postponed until the child is ideally 5 years old.

A decision to implant a child between the ages of 3 and 5 years is made on a case-by-case basis with the family within a multidisciplinary team.

Introduction

In 1965, Branemark first introduced osseointegrated titanium dental implants¹. These proved to be a great success. Tjellström and his team reported the first bone-anchored hearing aids.^{2,3} Today, the Bone anchored hearing device (BAHD) is a well-described and accepted form of auditory rehabilitation.

In the adult population, the technique is commonly performed as a one-stage procedure often using local anaesthesia. In the paediatric population many centres prefer general anaesthesia and two-stage surgery, typically allowing a period of 12 weeks between stages.

Many children who benefit from a BAHD have a significant medical or syndromic history, which may influence the morbidity of the procedure.^{4,5} Furthermore, there are issues of soft tissue and skull thickness to consider. These vary greatly in young children and may significantly affect the surgical procedure itself as well as any subsequent complication rates.

Finally, when is the best time to implant a young child? This is always a difficult question. The aim of the clinician and audiologist is to restore hearing as soon as possible in order for the child to achieve their full potential. This must be balanced against the risk of surgical morbidity.

The Birmingham Paediatric BAHD programme began in 1992. In the first decade of the programme, it was not uncommon for children as young as 2 years of age were to be implanted. In more recent years, following the introduction of sound processors on soft headbands, the age for implantation has increased.

This study evaluates the morbidity of BAHD surgery in these very young children. Our aim was to evaluate the morbidity of bone anchored hearing aid surgery in children below the age of 5 years over the course of the BAHD programme.

Patients and methods

This was a retrospective case analysis of all children below the age of 5 years implanted with an osseointegrated fixture for a BAHD. The records for this study spanned an 18-year period (1992 -2010). As with any retrospective study, our data was collected from departmental databases and medical records.

Many children in this study had previously worn conventional hearing aids and the majority of children had undergone insertion of ventilation tubes with little success.

Parents and caregivers needed time for consideration of all the treatment options available. Typically, all patients had a trial of a bone conductor aid or Baha® softband™ and a minimum of two outpatient clinic appointments for discussion about the BAHD before consenting for surgery.

No children with a fixture retained aural prosthesis were included in this study. Funding for BAHD in the National Health Service has changed significantly over recent years and it is now accepted practice for children to be offered bilateral BAHD if they have a bilateral hearing loss. Simultaneous bilateral implantation began in 2008.

All the children underwent a two-stage procedure under general anaesthesia. Stage one involved the implantation of a fixture and a 'sleeper' fixture was common practice. The side of implantation was based upon the patient and surgeon's preference as well as cochlear function. Typically the second stage procedure, including soft tissue reduction and abutment placement was carried out 12 weeks later.

In the early stages of the Birmingham BAHD programme a local free split thickness graft was used at the second stage. In later years this was modified to a pedicled split thickness graft using the Cochlear dermatome®.

All children had their first post-operative visit on day 5 or 6. The follow-up was tailored to the individual clinical needs of the child. As a matter of routine, all children had one postoperative visit with the clinicians to assess the peri-abutment soft tissues at their BAHD site. More frequent visits were arranged if necessary. All patients were closely followed up in the interim period by a dedicated BAHD nurse practitioner who had many years of experience in dealing with BAHD wounds.

All records between 1992 and 2010 were evaluated by two clinicians (R.B. and A-L.M). The outcome measures included patient demographics, indication for BAHD, surgical procedures, complications and finally the number of children who are still current BAHD wearers.

Results

There were a total of 53 children aged 5 years or under at the time of implantation. The age range was 2 years to 4 years 11 months (mean 3yrs, 5mths). See figure 1.

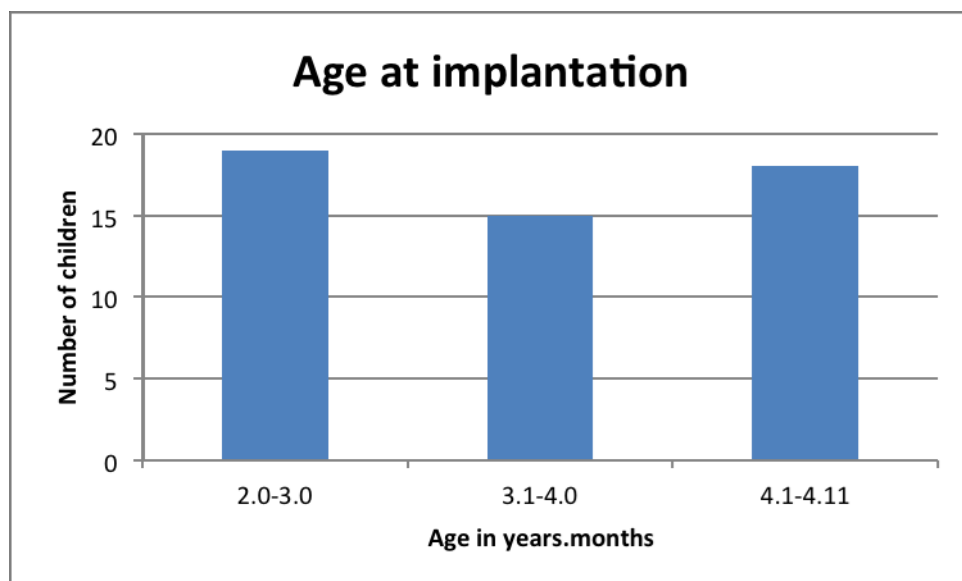


Figure 1. Age distribution at the time of implantation

The male: female ratio was similar (27M: 26F).

32/53 (60%) of the children had a significant medical history. See table 1.

Table 1. Co-morbidity identified in children below the age of 5 years who were fitted with a BAHD

Co-morbidity	Number of cases
Treacher Collins Syndrome	13
Chromosomal abnormalities	4
Hemifacial microsomia/Goldenhar syndrome	4
Nager Syndrome	1
Branchio-Oto-Renal Syndrome	2
Dubowitz Syndrome	1
Pierre Robin Sequence	1
Antley Bixler Syndrome	1
Binder's Syndrome	1
Down Syndrome	3
Epilepsy	1
No co-morbidity	21

The most common indications for BAHD implantation are illustrated in table 2.

Table 2. Indication for BAHD Insertion.

External ear malformation	Number of cases
Treacher Collins Syndrome	13
Isolated bilateral atresia	18
Various chromosomal abnormalities	4
Hemifacial microsomia/ Goldenhars Syndrome	4
Nager Syndrome	1
Dubowitz Syndrome	1
Pierre Robin Sequence	1
Antley Bixler Syndrome	1
Binder's Syndrome	1
Chronic Suppurative Otitis Media/OME	Number of cases
Treacher Collins Syndrome	2
Isolated bilateral atresia	2
Branchio-Oto-Renal Syndrome	2
Down Syndrome	3

In total there were 53 children with 62 ears implanted

Side of Implantation: During the study period, 44 children had a unilateral BAHD (28 Left, 16 Right) and an additional 9 children had bilateral implantation.

Sleeper fixtures: The majority of children had a fixture and a second 'sleeper fixture' implanted, but in 3 children it was impossible to implant the 'sleeper' due to insufficient thickness of the calvarial bone.

Implanted Fixtures: 3mm fixtures were used in 50/53 (94%) children. The remaining 3/53 (6%) children had a combination of 3mm and 4mm fixtures.

Bone thickness: In 30/53 (57%) children, more than one attempt was made to find suitable thickness of bone at the time of surgery. This involved careful exploration at the time of surgery (by drilling pilot holes) to locate the best location for the implant.

The most common operative findings included thin calvarial bone and exposed dura. See table 3.

Table 3. Operative findings

Operative findings	Number of children
Thin calverial bone	17
Exposed dura	16
Gore-Tex® membrane used	9
Sigmoid sinus bleeding	1
Failure to implant	1

Where thin bone was encountered, the dura was depressed with a 3mm fixture. A Gore-Tex® membrane was placed under the fixture flange to promote osseointegration in 9/53 (17%) cases, a technique described by Granström et al(6).

The time between the first and second stage varied and appeared to be based on the operative findings. This ranged from 12 to 51 weeks (mean 20 weeks).

Peri-abutment skin reactions: 26/53 (49%) children had a significant skin reaction around the abutment following the second stage of surgery. We defined a “significant skin reaction” as any wound that required more than three return visits to the outpatient clinic, repeated topical medications, and/or surgical reduction.

3 children 3/53 (6%) had complete graft failure. These wounds healed by secondary intention and did not require any further surgery.

In total 17/53 (32%) children required surgical skin reduction as a revision procedure. Histological samples sent at the time of surgery demonstrated a foreign body granulomatous reaction to keratin. There was one child with Treacher Collins Syndrome who was implanted at the age of 4. He had bone growth around the fixture site at the age of 15, but this was overcome with the use of a longer 12mm abutment.

Fixture failure: 21 children out of the 53 suffered a fixture loss (39.6%) and of the 62 ears implanted, 21 had a fixture loss giving an overall fixture loss rate of 33.8%. Six failures were as a direct result of trauma. All but one of the fixtures lost were 3mm. All of the non-trauma related fixture losses occurred within the first two years of implantation. See table 4.

Table 4. Fixture failure Group

Case	Age (y.m)	Gender	Number of fixture losses	Medical history
1	3.7	M	3	Isolated bilateral atresia
2	3.11	F	3	Isolated bilateral atresia
3	2.8	M	3	Isolated bilateral atresia
4	4.2	F	3	Branchio-Oto-Renal Syndrome
5	4.11	F	2	Isolated bilateral atresia
6	2.9	M	2	Treacher Collins Syndrome
7	4.6	F	2	Isolated bilateral atresia
8	4.2	F	2	No medical co-morbidity
9	3.8	M	1	Chromosome 18 deletion (de Grouchy Syndrome)
10	2.2	F	1	Treacher Collins Syndrome
11	4.3	M	1	Goldenhar Syndrome
12	4.11	F	1	Isolated bilateral atresia
13	3.9	F	1	Isolated bilateral atresia
14	4.8	M	1	Goldenhar Syndrome
15	2.3	F	1	Treacher Collins Syndrome
16	2.5	F	1	Isolated bilateral atresia
17	2.8	M	1	Isolated bilateral atresia
18	4.9	M	1	No medical co-morbidity
19	3.6	M	1	Isolated bilateral atresia
20	4.8	M	1	Chromosome 4 anomaly
21	2.4	M	1	Isolated bilateral atresia

Evaluation of the fixture loss (FL) and non-fixture loss (NFL) groups was performed to establish if there were any potential risk factors for the soft tissue complications.

- a) In the NFL group, 23 out of the 32 children (71.9%) had a recognised medical comorbidity.
- b) In the FL group 8/21 (38.1%) had a recognised medical co morbidity.

None of these co morbidities were recognised to have an increased risk of wound infection although those associated with learning difficulties were at greater risk of problems with maintenance of the health of the peri-abutment soft tissues.

It was interesting to note that of the 8 children who had multiple fixture loss, (MFL) only one had a significant medical co-morbidity; the others had isolated atresia with no other medical history or risk factors of note.

Interestingly, the mean age at implantation was similar in all three groups.

The time between surgical stages ranged from 12-51 weeks (mean 20 wks.). All the fixture failures and significant soft tissue problems were encountered within the first two years.

Current BAHD wear/ “Success”

Currently all 53 (100%) children from this study are active BAHD wearers.

Discussion

BAHD in children is a well described and well accepted form of auditory rehabilitation.^{5,7-9}

It can be used in those children who cannot tolerate conventional hearing aids, persistent middle ear problems, bilateral congenital ear malformations and more recently children with severe learning/behavioural difficulties.¹⁰⁻¹²

Compared to the adult literature on BAHD there are far fewer publications regarding BAHD in the very young. Most of the literature includes children of all ages. Of the papers published on results in children over the last 8 years, only one other paper from the group in Toronto specifically targets this very young group.¹³

See table 5

Parents and carers need time for consideration of all the treatment options available. There is sufficient evidence to show that the Baha® softband™ outperforms the traditional bone conduction aid. Christensen et al found that in children with bilateral conductive hearing loss, the Baha® softband™ provided statistically significant functional gain and threshold benefit compared with traditional bone conduction devices.¹⁴

Earlier studies have shown that children implanted at 1 year of age, were in fact children of parents who themselves were BAHD wearers.³ These parents required less time for their decision-making.

Table 5 - an overview of BAHD outcomes in children

Study	Year	N	Mean age (y)	Surgical technique	Fixture length	Number of stages	Soft tissue reduction (%)	Fixture loss %
Current Study	2013	53	3.5	Split skin graft	3mm in 94%	2	32	33.8
Lanis et al. Stockholm	2013	34	6.5	23 skin thinning, 10 nonthinning	3mm in 94%	1 and 2	10 thinning 0 nonthinning	21.7 thinning 10 nonthinning
de Wolf et al. Nijmegen	2008	93 (12 under 5)	9.3	Linear incision	3mm in 24%	2 in under 10ys	9.4	16.3
Davids et al. Toronto ¹²	2007	20	3.2	Not mentioned	3mm in 100%	2	15	10
Lloyd et al. London	2007	71	8.7	Various	3mm in 73%	2	25.9	28
Yellon. Pittsburgh	2007	14	5.8	Split skin graft	Not mentioned	2	28.6	14.3
Priwin et al. Stockholm	2005	41	8.4	Thinned soft tissue flap	3mm in 45.5%	2	17.1	9.1

No preoperative imaging was undertaken in any child. The assessment of skull thickness is not best appreciated by conventional radiology. The skull contour and position of the external ear (if present) primarily determines the position of the BAHD. Computerised tomography in this young age group may require sedation or general anaesthesia and exposes the child to a large radiation dose.^{4,5} In addition to this, it is difficult to mark the optimal radiological site in order for it to be utilised on the operating table. In our experience, simply careful exploration at the time of surgery and the drilling of pilot holes is sufficient to determine the site of BAHD. In addition to this, with the advent of better quality sound processors mounted on headbands, surgery in children can now be successfully delayed without any compromise in hearing until the age of 4-5 years when calvarial thickness is more favourable and morbidity is less. Perhaps as the use of cone beam computed tomography becomes more widespread it may be an additional tool used in the preoperative planning of a BAHD due to the lower radiation dose.

Patient details

In total 53 patients were identified and all 53 records were analysed. Of these, one set had no operative data, and two other sets of records were incomplete. The details were retrieved from the electronic operating room database and old operating ledgers.

The indications for BAHD are shown in table 2. The Birmingham Children's Hospital is a large paediatric tertiary referral centre; and this accounts for the high proportion of unusual and rare medical conditions in this study. This is similar to those studies from other large specialist Children's hospitals.^{10,13,15-19}

Surgical procedure

When is the ideal time to implant a young child? There has been much debate over this issue in recent years, and most of the paediatric BAHD literature reports implantation in children aged 4 years and above.^{20,21} Tjellström et al have recommended that age 3 years and above is ideal since the child should then have suitable skull thickness.²¹ The surgical technique has been described extensively by Tjellström.²²

Despite recent reports describing single stage procedures in children, all the children under five years of age in our centre underwent a two stage procedure under general anaesthesia.²³ Our programme still employs this two-stage technique in these young children because of the increased risk of trauma, and the increase risk of failure of primary osseointegration.

The increased morbidity of BAHD in these very young children has to be balanced against the effects of delaying maximum auditory habilitation. There is evidence that a mild conductive hearing loss can affect language acquisition and development in children with no coexisting learning difficulties.¹⁸ Mani et al recently described a two stage BAHD procedure with just one general anaesthetic in children.²⁴ The BAHD team in Glasgow performs a single stage procedure in children as young as three years of age and reported a very low rate of early complications and fixture failure.²³

In the early years of the Birmingham BAHD programme, a local free split thickness graft was used at the second stage. In later years this was modified to a pedicled split thickness graft using the Cochlear dermatome®. More recent literature has reported excellent results using a linear incision technique.¹⁰ It is interesting that recent studies have shown that local split thickness grafts were less likely to develop graft hypertrophy than the Wolfe grafts.¹⁶ Wolfe grafts were not used in the Birmingham programme. There are many other techniques described in the literature.²⁵ Most recently a study from Sweden has reported a lower fixture failure and surgical soft tissue reduction rate in children with a non skin thinning technique, although long term results are still awaited.²⁶

The most common difficulty encountered at the time of surgery was thin calvarial bone however the low number of documented intra-operative findings most likely reflect the lack of details recorded in the operative journal. (Table 4.) Since 17/53 (%) of these children were below the age of 3 years it is probable that dura would have been encountered in these cases.

Dura and sigmoid sinus exposure during BAHD surgery have been reported to be as high as 70% in the paediatric population⁶ In our centre, common practice was to explore multiple sites on the temporal bone (at the time of surgery in one sitting) when inadequate bone thickness was encountered. The Gore-Tex® augmentation technique for this situation is well described.³ The use of a Gore-Tex® membrane under the fixture flange was documented in 9 of the children in our study.

Time between surgical stages

Typically three months between surgical stages is advocated. The long period between single stage surgery and loading of the sound processor has been a limitation until recently.²⁷⁻²⁹ Wazen et al²⁹ demonstrated no significant increase in fixture failure with a healing time of just 6.5 weeks between surgery and loading in adults. Furthermore, Zeitler et al³⁰ have demonstrated that in single stage bone anchored implantation in the adult population, there were no reported cases of failure to osseointegrate in patients loaded as early as 6 weeks post operatively. Dun et al reports equally good results in adults for extrusion rates and skin reactions for loading times between 3-5 weeks compared to groups with loading times at 6-8 weeks, 9-11 weeks and for over 12 weeks.³¹

Recent evidence is emerging that a newly designed titanium coated rough surfaced fixture (BI300) is reducing the healing time between surgical stages. This is allowing successful loading of the sound processor as early as two weeks with no increase in morbidity. This fixture has a wider diameter and conical structure, with smaller outer threads. The surface has been treated with titanium oxide. It is not known if the change in shape of the implant and/ or the change in the surface texture is influencing these results³¹ The dental literature has demonstrated successful osseointegration with loading times ranging from the immediate postoperative period up to 8 weeks.³²

Davids et al¹³ has described the placement of a 3mm fixture flush with the outer bone table (dura depressed by the fixture) then a longer period of healing time before the second surgical stage. They found this reduced the need for multiple pilot holes. Their osseointegration rate was 90% in a similar group of very young children.

Comparing results from different centres is difficult as they represent very different population groups. This in turn may explain the differences in paediatric BAHA complications.¹⁰

In our study there was an average of 20 weeks healing time (ranging from 12-51 weeks). It is worth noting that there was one child with a very long healing time of 51 weeks and this was due to social reasons. In fact the second longest healing time was 20 weeks. Interestingly the Toronto group(12) did have a longer healing time with a mean interval of 7.7 months between the first and second stage of surgery compared to our mean inter-stage healing time of 20 weeks (5 months).

Fixtures

The fixture failure rate in our study was 33.8%. This is higher than that reported in adults^{33,34} and is in keeping with literature from the Great Ormond Street BAHA team.¹⁴ Their fixture failure rate was 26% and their soft tissue reaction rate was 37% with 9% lasting more than 6 months. They identified young age at implantation to be associated with an adverse outcome.³³

All but one of the lost fixtures in our series was 3mm. It is interesting that some previous studies have reported an increased failure rate with the shorter fixtures.³³ We were unable to implant one child in our study. She had a rare autosomal recessive condition noted to involve delayed bone maturation. She has since been successfully implanted and is currently wearing her sound processor with good results.³⁵

Were there any obvious risk factors for fixture failures in these young children?

We hypothesised that the fixture failure rate might be higher in very young children with significant medical problems, however, there appeared to be no significant increase in the fixture failure rate in those children aged below 3 years. Similarly there seemed to be no increased fixture failure rate in those patients with medical co-morbidity. No obvious potential factors were eventually identified in this group of young children. In a previous study performed at Great Ormond Street Hospital¹⁶ describes 26% of their children experiencing fixture loss(es) at some point. This study included children with an age range of 3.6 – 17.2 years, and furthermore, they reported that there were increased complications associated with younger age at implantation.

Soft Tissue complications

The percentage of young children with a skin reaction was 49% with 32% requiring surgery. This is similar to other reports in the paediatric BAHD literature.^{34,36}

Although Holger's Classification³⁷ was first described in 1987, it was not routinely used in Birmingham until after 2001. See table 6.

Table 6. The classification of skin reaction around the abutment described by Holger KM³⁷

Score	Soft tissue description
0	No irritation
1	Slight redness
2	Red, slightly moist, no granulations
3	Red, moist, with granulations
4	Loss/removal of skin penetrating implant. Revision required

Early records pertaining to soft tissue complications were very subjective and difficult to standardise. Photographic documentation has been invaluable in the assessment of wounds.

One specialist Baha nurse who has many years of experience carried out the soft tissue assessment. A significant soft tissue reaction was defined as any wound that required more than three return visits to the outpatient clinic, repeated topical medications, and/or surgical reduction.

Daivids et al¹³ reported a low fixture failure rate of 10% in children under 5 years, all of which were as a result of trauma. Their soft tissue complication rate was also low and only 15% required revision skin surgery. This was in comparison to our surgical skin reduction rate of 32% in a similar group of children.

The Baha® softband™

In 2002, the Baha® softband™ was introduced. Hol et al demonstrated the Baha® softband™ provided improved audiological benefit when compared to conventional bone conduction aids in children with congenital bilateral aural atresia; thus providing a valid treatment option for young children.^{14,38} It has also been demonstrated to provide a well tolerated and non-invasive method of managing conductive hearing loss as a result of otitis media with effusion in young children.^{39,40}

A bone conductor headband is currently the treatment of choice for children under 3 years of age in Birmingham. This provides all the benefits of BAHD delivered in

an acceptable and well-tolerated form for these children without compromising their speech, language and general cognitive development. It also allows time for physical growth especially skull thickness. In our experience, the bone conductor has been so popular with very young children that families and carers are keen to proceed to surgery as soon as the child reaches the age of three years. There is increasing parental pressure for younger implantation.

For children aged between 3 and 5 years, the treatment decision is made after discussion between the patient, family and the BAHD multidisciplinary team.

Finally trauma is a challenge to any paediatric BAHD team. In our study 6 children lost their fixtures as a direct result of a traumatic injury. One case from this series sustained an intracranial intrusion of both the fixture and abutment.⁴¹ The majority of soft tissue and fixture complications in our study occurred within two years of the surgery however, trauma occurred at any time. The BAHD multidisciplinary team has a role in the care of their patients long after the surgical procedure is complete.

Conclusions

The use of the BAHD in children under 5 years of age has previously been controversial.

In the Birmingham series, there was an increase in morbidity in this young patient group compared with adults and older children. The fixture failure rate was high as was revision surgery for significant skin reactions.

In 2002, the Baha® softband™ was introduced. Since then, in Birmingham, the treatment of choice for children less than 3 years of age is the Baha® softband™ or alternative bone conduction headbands. They are safe, non-invasive and well accepted by young children allowing development of speech and language. Since there is a recognized increase in morbidity associated with BAHD in very young children, our policy is to ideally reserve surgery until the child is older. For children aged between 3 and 5 years, the treatment decision is made after discussion between the patient, family and the BAHD multidisciplinary team.

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Chapter 3

Quality of Life outcomes following Bone Anchored Hearing Device (BAHD) Surgery in Children with Single Sided Sensorineural Deafness.

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Abstract

Objective: To report our experience in a series of children with single sided sensorineural deafness where a bone-anchored hearing device (BAHD) was used for auditory rehabilitation.

Study Design: Retrospective case review.

Setting: Tertiary referral centre.

Patients: Eight children (4 male & 4 female) who had BAHD surgery for single sided sensorineural deafness between 2007-2010.

Intervention(s): Bone-anchored hearing device was used for auditory rehabilitation.

Main Outcome Measure(s): Glasgow Children's Benefit Inventory (GCBI), Single Sided Deafness (SSD) Questionnaire and change in health benefit scores (visual analogue scale).

Results: All but one of the children showed a positive GCBI score; the child that reported a negative score was due to low self-confidence and self-esteem issues secondary to bullying at school. The results of the SSD questionnaire were generally positive with a mean satisfaction score of the BAHD as 9/10. All the children had an improvement in health benefit.

Conclusions: Our findings add further evidence to support patient perceived benefit of a BAHD in single sided sensorineural deafness in the paediatric population.

Introduction

Single sided deafness (unilateral profound sensorineural hearing loss) has been reported to have an incidence between 0.1 to 3% in the paediatric population.^{1,2}

The cause of the hearing loss may be congenital or acquired. Historically, treatment options ranged from no intervention (especially if the contralateral ear had normal hearing thresholds) to FM amplification systems and contralateral

routing of signal (CROS) aids; these latter devices having limited user compliance.^{3,4}

In adults with single sided deafness, studies have shown both objective and subjective improvement in audiologic metrics with a bone anchored hearing device (BAHD) when compared to unaided conditions.^{5,6,7} Studies have shown that adults with single sided deafness treated with a BAHD do have an improved quality of life^{5,8} however this benefit may not be as great as other patient groups (e.g. conductive or mixed hearing loss).⁹ Compared to the adult population, there is less experience of the outcome of using a BAHD in children with single sided deafness.

The aim of this paper is to report our experience in a series of children with single sided sensorineural deafness where a BAHD was used for auditory rehabilitation.

Material and Methods

Eight children (4 male & 4 female) had BAHD surgery for single sided sensorineural deafness between 2007-2010.

All the children had been assessed by a community paediatrician and been fully investigated for the aetiology of the hearing loss.

The paediatric audiology department had referred these children to the otolaryngology department on the request of parents reflecting educational concerns resulting from their child's hearing.

All the children were assessed in the multi-disciplinary BAHD clinic and underwent a trial of using a bone conductor on a headband for a minimum of 3 months.

The parents/carers of the children were asked to complete a Glasgow Children's Benefit Index (GCBI) and a Single Sided Deafness (SSD) Questionnaire after a minimum of 6 months of BAHD usage.

The GCBI is a subjective child orientated post-interventional questionnaire especially developed to evaluate any paediatric otolaryngology surgery and therapy.¹⁰ The SSD questionnaire was specifically designed to be administered after BAHD implantation for single sided deafness; the questionnaire was first used in clinical study in 2003¹¹. It is based upon a questionnaire developed by Entific Medical Systems, which was published in the product's audiology manual.¹² We also asked the parents to mark on a 10cm visual analogue scale their child's health status before and after the BAHD.

Results

Clinical data

Four children had congenital hearing loss and four children had an acquired hearing loss. Patient 5 had sustained a skull base fracture following trauma at the age of 9 years old. Patient 6 had meningitis aged 3 months old. In the remaining group of children, no aetiology for the hearing loss had been identified.

The age at which surgery was performed ranged from 7.5 years old to 12.2 years old. The patient demographics are summarised in table 1.

Table 1: Patient Demographics

Patient	Cause of unilateral hearing loss	Ear with single sided deafness	Age of referral for BAHD	Age of BAHD surgery	Year of surgery	Length of follow up (months)
1	Congenital - unknown	Left	5	9.5	2009	27
2	Congenital - unknown	Left	9.8	10.7	2007	56
3	Congenital - unknown	Left	6.2	7.5	2010	16
4	Congenital - unknown	Right	10.4	11	2009	30
5	Acquired - skull base fracture	Right	9	12.2	2007	52
6	Acquired - meningitis	Left	9.6	10.1	2008	41
7	Acquired - unknown	Left	8.2	8.9	2009	30
8	Acquired - unknown	Right	6.6	8.3	2010	21

None of the children had any significant co-morbidities. They all had normal or near normal air conduction hearing thresholds in the contralateral ear (table 2). Patient 6 had the worst air conduction thresholds in the contralateral ear [mean air conduction threshold of 23.3dB (0.5, 1 and 2KHz)] which may have been related to the episode of meningitis that caused SSD in the contralateral ear.

All the children underwent a two-stage procedure with a minimum period of 3 months between the first and second stages of surgery. A “skin dermatome” technique was used in all cases. A 5.5mm abutment was used on 4mm fixture in

all the children. There were no postoperative skin complications with no cases of implant extrusion (follow up 16-56 months). All the children used a BAHA® Intenso processor on their abutment.

Table 2. Air conduction thresholds in the better hearing ear

Pt	250Hz	500Hz	1kHz	2KHz	4KHz	8kHz
1	25	20	5	-5	-5	0
2	10	5	5	-5	0	-10
3	-	20	20	5	10	-
4	-	5	0	0	0	-
5	20	20	15	15	5	25
6	35	25	25	20	20	10
7	15	15	5	5	5	-
8	5	5	5	-5	-10	-

All patients had a profound sensorineural hearing loss in the poorer hearing ear

Questionnaire results

GCBI

The GCBI scores ranged from -6.3 to 68 (figure 1) with a median of +47.5. All patients but one child (patient 5) showed an improvement in GCBI results. An analysis of the subcomponents making up the GCBI score of patient 5 showed that the negative scores were within the “emotional” subcomponent of the GCBI whereas the “health”, “learning” and “vitality” subcomponents were neutral or positive. On further questioning, patient 5 had confidence and self-image issues secondary to bullying at school.

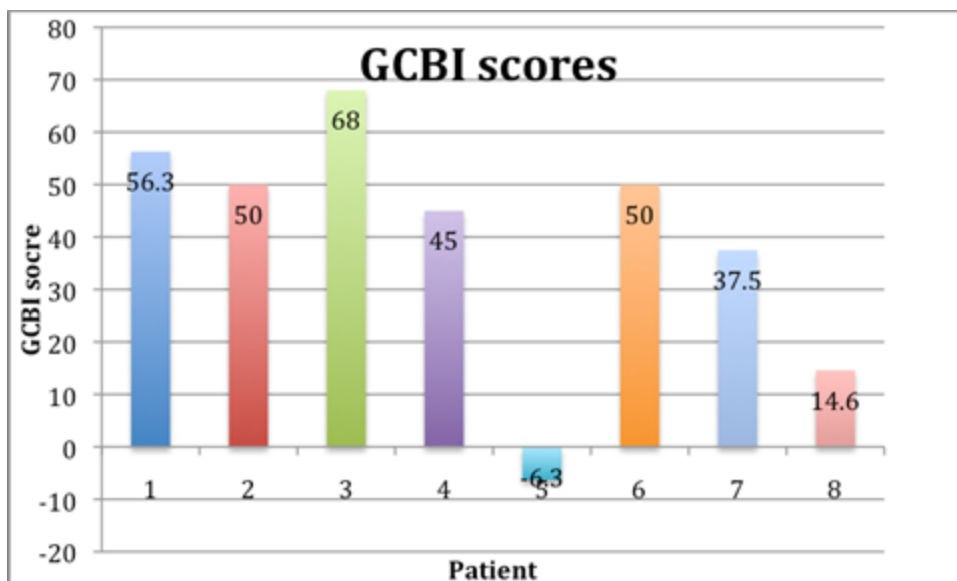
SSD questionnaire results.

The majority of children (6/8) used their BAHD for 7days/week and 7/8 children used the BAHD for more than eight hours a day (figures 2 and 3)

The average satisfaction score was 9/10 with the BAHD; 5/8 thought the quality of life had improved.

There was a subjective benefit whilst using the device one-to-one in half the children; the other half did not report a difference.

Using the device within a group, all but one of the children felt the BAHD was beneficial. There was perceived benefit by the majority of children when using the device to listen to music, TV or radio. There was also a perceived benefit of the BAHD by the vast majority of children when someone spoke to them on their deaf side in a daily commonplace scenario (at the dinner table). Results are summarised in figure 4.



GCBI - Glasgow Children's Benefit Inventory

Figure 1. Glasgow Children's Benefit Inventory Scores

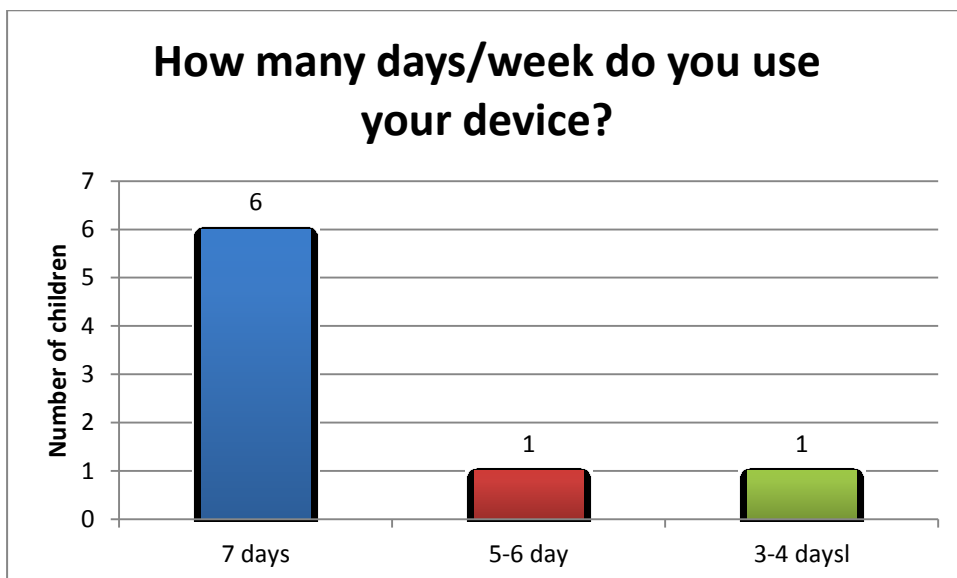


Figure 2. Frequency of device usage

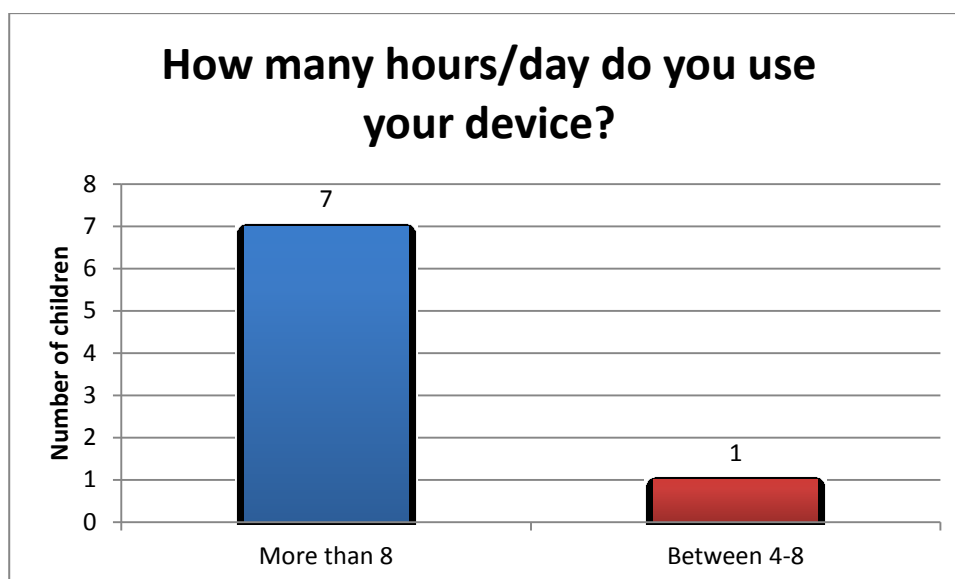


Figure 3. Intensity of device usage

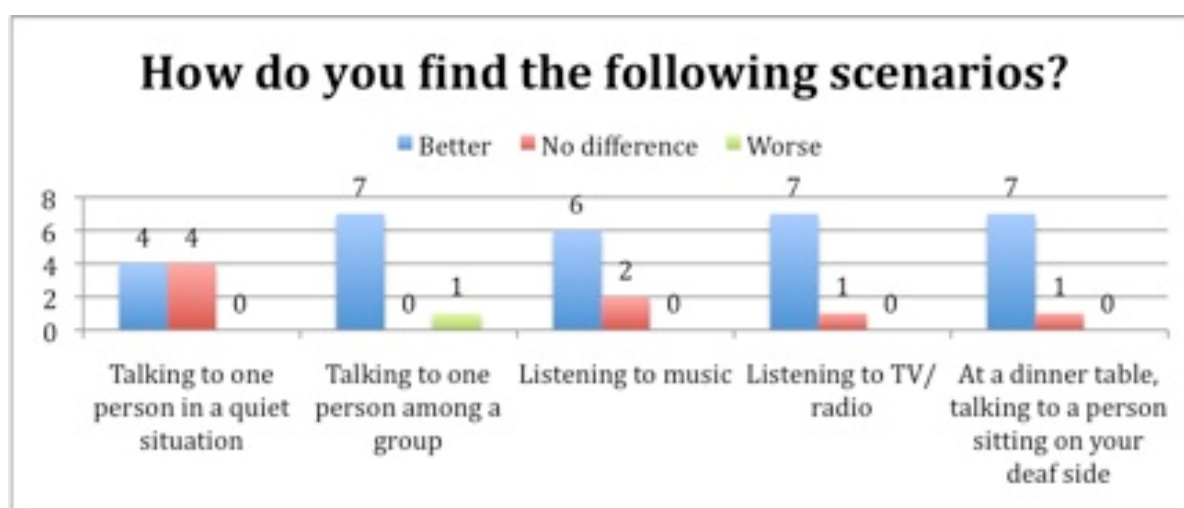


Figure 4. Practical benefit of the device

Change in health status

For all the children, parents reported an improvement in health status following surgery (figure 5). The median change in health status following BAHD surgery was +4 (range 3-4.9)

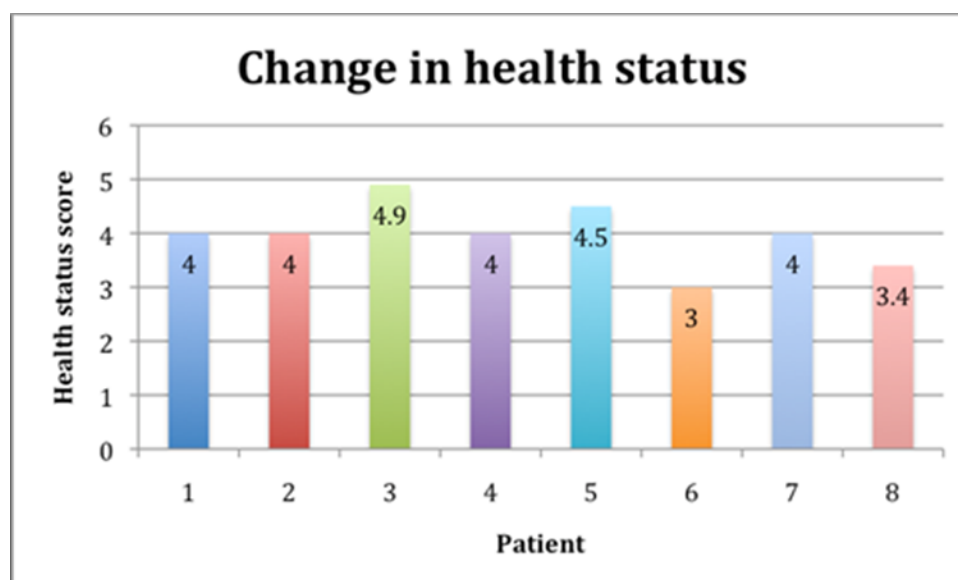


Figure 5. Change in health status

Discussion

Our paper suggests that an BAHD does improve the quality of life in children with SSD and adds to the sparse literature on this topic.

Christensen et al reviewed 23 children (age range 6-19) that had an BAHD for single sided deafness at their institution.¹³ They assessed outcome using two questionnaires - Hearing in Noise Test (HINT) and Children's Home Inventory for Listening Difficulties (CHILD). Scores, before and after surgery, showed noticeable improvements in both these outcome measures with the BAHD. The authors had previously shown similar results in a pilot study of three teenagers with single sided deafness.¹⁴ Gluth et al also demonstrated improvement in HINT and CHILD scores post BAHD surgery in a series of three teenagers with single sided deafness.⁸

In our series of 8 children, we used the GCBI and SSD questionnaire to assess benefit. All but one of the children showed a positive GCBI score; the child that reported a negative score was due to low self-confidence and self-esteem issues secondary to bullying at school.

Compared to adults with SSD treated with an BAHD, the results in our paediatric

population are more favourable. The median GCBI score in our cohort of children was +47.5. Martin et al reported a median GBI score of +11 in 42 SSD BAHD adult users.⁹

The GCBI scores from this study are also comparable to the benefit obtained by adults with bilateral devices for conductive hearing loss. Ho et al reported GBI score of +38 with the use of bilateral BAHDs.¹⁵

Over half of the children in this paper had a GCBI score equal or above +50. The magnitude of the positive GCBI scores reflects comparably to other paediatric BAHD indications. A series of 84 children from our institution showed a mean GCBI score of +54 following BAHD surgery for chronic suppurative otitis media, atresia or syndrome-related conductive hearing loss (Down's/Treacher Collins/Goldenhar syndrome).¹⁶

The results of the SSD questionnaire were generally positive with a mean satisfaction score of the BAHD as 9/10.

Admittedly, our series of 8 children is relatively small. They had all been referred because of parental concern with their children's educational development therefore it could be argued that selection bias was introduced as the questionnaire results were from a "motivated" parent/child population. In addition, the GCBI and SSD questionnaires assess subject satisfaction therefore they explore whether the device meets or exceeds expectations; the results can be affected by positive bias particularly if respondents feel grateful for intervention even if it is not technically or objectively successful.¹⁷

A recent study has also found there is a wide variation between individuals (both adults and children) in the transcranial attenuation of bone-conducted sound therefore this may also influence subjective benefit of a BAHD.¹⁸

In summary, our findings add further evidence to support patient perceived benefit of a BAHD in single sided sensorineural deafness in the paediatric population. The magnitude of benefit can be comparable to the more established indications for a paediatric BAHD. Further work is required in a larger series of children to investigate if this benefit is statistically significant.

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Chapter 4

Bone anchored hearing devices in children with unilateral conductive hearing loss: A patient/ carer perspective

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Abstract

Objective: To determine the outcome of a bone anchored hearing device in children with a unilateral conductive hearing loss.

Methods: A retrospective case note analysis in a tertiary referral paediatric hospital. A total of 17 consecutive paediatric patients with a unilateral conductive hearing loss were fitted with a bone anchored hearing device between 2005 and 2010.

Results: The average age of bone anchored hearing device fitting ranged from 6 years, 3 months to 16 years. Average age at fitting was 10 years, 6 months.

Qualitative subjective outcome measures demonstrated benefit.

The vast majority of patients reported improved social and physical functioning as well as improved quality of life.

All 17 patients are currently using their bone anchored hearing device on a daily basis after a follow up of 6 months.

Conclusions: This study has shown improved quality of life in children with unilateral hearing loss after receiving their bone anchored hearing device. There was a high patient satisfaction and improvement in health status reported by children/carers. Bone anchored hearing devices have an important role in the management of children with symptomatic unilateral hearing loss. Perhaps earlier consideration of a bone anchored hearing device would be appropriate in selected cases.

Introduction

In 1987, the first semi-implantable bone conduction devices became commercially available.¹⁻³ Initially when bone anchored hearing devices (BAHD) were first introduced, they were used primarily for those patients in whom it was not possible to wear conventional air conduction aids. This typically included patients with chronic middle and external ear disease, congenital malformations of the external auditory meatus and pinna. Recent years has seen the indications for a BAHD expand.^{4,5}

The percutaneous BAHD is currently a well recognised and very effective method of rehabilitation for patients with both unilateral and bilateral conductive and mixed hearing loss.^{6,7}

It is well known that a bilateral hearing loss in children can cause problems with speech and language development, and if not recognised early can ultimately affect educational achievements and have an impact on the child's behaviour.

The magnitude of morbidity imposed upon a child with a unilateral hearing loss is much less understood. Some children appear to perform well and have no apparent disadvantage from their unilateral hearing loss (UHL), yet others are more handicapped by their UHL. In 2004 Lieu et al⁸ showed that there was a significant proportion of children with a UHL that had educational or behavioural problems at school when compared to their normal hearing peers. Christensen et al have shown that children with UHL find benefit with BAHD both on audiological testing and patient satisfaction questionnaire⁹ and Priwin et al has shown that the fitting of a BAHD in children with unilateral conductive hearing loss (UCHL) leads to improved speech recognition in noise, but less favourable improvement in sound localization.¹⁰ In addition to this, Kunst et al found that some patients with a UCHL had such good unaided directional hearing and speech in noise scores that aided testing with the BAHD did not confer significant overall improvement. Despite this the compliance with BAHD use was very high in this group of patients suggesting patient benefit.¹¹

Are there any risk factors to predict the children that will struggle? How should these children be treated? Should they be ignored until there is an evident educational need, should they all be aided or is there a compromise?

This paper aims to evaluate the impact that BAHD has on the quality of life in children with symptomatic unilateral conductive hearing loss (UCHL).

Patients and methods

All children fitted with a BAHD for UCHL between 2005 and 2010 were identified from a departmental database. There were 17 in total. A retrospective case note review was undertaken. As part of their on going follow up, all these children and/or carers completed a Glasgow Children's Hearing Aid Benefit Profile (GCBI) and a Single Sided Deafness (SSD) Questionnaire. They also used a visual analogue scale to indicate their perceived health status before and after their BAHD which

has been used in other published quality of life papers and has been shown to correlate with GCBI score.^{5,12,14}

The GCBI is a validated subjective child orientated post-interventional questionnaire designed to evaluate any paediatric otolaryngology intervention.¹⁵ It consists of 24 questions based upon a five point Likert scale. A score of +2 shows a maximal positive change and a score of -2 a maximal negative change. The sum total score is divided by 24 and multiplied by 50 to give a score ranging from +100 to -100 depicting a positive or negative change. Specific questions in the inventory relate to emotion, physical health, learning and vitality. See Appendix 1.

The SSD questionnaire was specifically designed to be administered after BAHD implantation for single sided deafness. It is designed to evaluate the number of hours and days that the BAHD is used and also to evaluate the benefit in various social situations. The questionnaire was first used in clinical study in 2003¹⁶ and it is based upon a questionnaire developed by Entific Medical Systems, which was published in the product's audiology manual.¹⁷ See Appendix 2.

The 10cm visual analogue scale was used to determine the change in health status before and after the BAHD where a positive change represents benefit to the overall health of the child and negative change represents deterioration.

Results

Age at referral

A total of 17 consecutive paediatric patients with UCHL were fitted with a BAHD between 2005 and 2010. The age at referral ranged from 3 months to 14 years with an average of 7 years and 6 months. The age at BAHD fitting ranged from 6 years, 3 months to 16 years, with an average age of 10 years, 6 months. The average age of referral for the children with congenital and acquired UCHL was 7 years, 2 months and 8 years, 2 months respectively. There was one child with a congenital UCHL that was fitted 8 years after the first referral to the otolaryngology clinic. She was seen in the audiology clinic six months prior to fitting but it was not clear in the records why there was such a long delay between the appointments. It was apparent from many of the records that the families had sought help from audiology because they firmly believed that their child had educational or speech and language difficulties as a result of their hearing impairment.

Aetiology of hearing loss

Congenital abnormalities accounted for 12/17 (71%) of the cases. The remaining 5/17 (29%) were acquired. See Tables 1 and 2.

Table 1. Congenital unilateral conductive hearing loss cases

Case	Aetiology	Affected side	Age of onset of deafness	Age at referral	4 tone average (0.5,1,2,4 kHz)		Age at BAHD fitting	Year of BAHD fitting
					L	R		
1	Microtia and atresia	L	Birth	7y	70	10	15y	2009
2	Hemifacial microsomia	L	Birth	3m	80	15	6y 3m	2008
3	Isolated unilateral bony ear canal atresia	R	Birth	7y	20	55	8y	2009
4	Hemifacial microsomia	L	Birth	10y	80	5	12y 5m	2009
5	Microtia and atresia	R	Birth	2y	20	70	7y 6m	2008
6	Hemifacial microsomia	L	Birth	10y	65	10	11y 9m	2006
7	Isolated unilateral bony ear canal atresia	L	Birth	8y	70	15	10y 3m	2008
8	Microtia and atresia	R	Birth	7y	0	65	9y 2m	2009
9	Microtia and atresia	L	Birth	9y	55	0	9y 3m	2003
10	Congenital ossicular malformation	R	Birth	7y	20	65	10y	2009
11	Microtia with atresia	R	Birth	11y	10	70	12y 8m	2006
12	Microtia with atresia	R	Birth	8y 2m	5	70	9y 8m	2010

Table 2. Acquired unilateral conductive hearing loss cases

Case	Aetiology	Affected Side	Age of Onset	Age at referral	4 tone average (0.5,1,2,4 kHz)		Age at BAHD fitting	Year of BAHD fitting
					L	R		
1	Chronic Suppurative Otitis media	L	4y	4y	60	20	9y 6m	2009
2	Chronic Suppurative Otitis media	R	8y	9y	5	50	13y 3m	2009
3	Chronic Suppurative Otitis media	L	?	5y	80	25	8y 3m	2008
4	Chronic Suppurative Otitis media	R	6y	9y 3m	10	45	10y 3m	2010
5	Chronic Suppurative Otitis media	R	?	14y	10	60	16y	2010

GCBI results

All but one child reported a positive change on the GCBI after having a BAHD for their UCHL. See Figure 1.

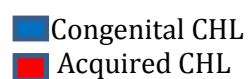
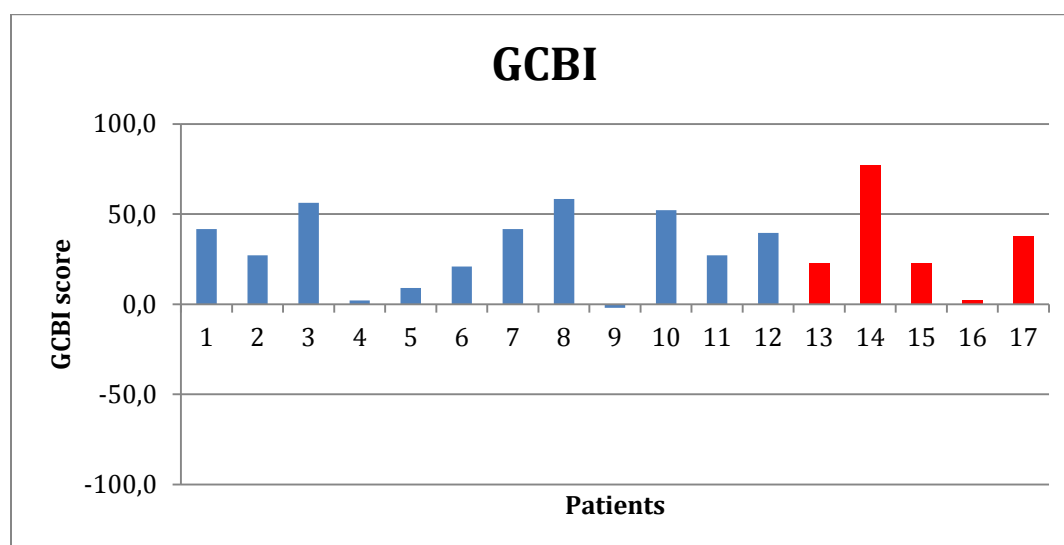


Figure 1. GCBI results

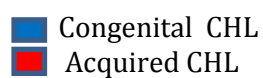
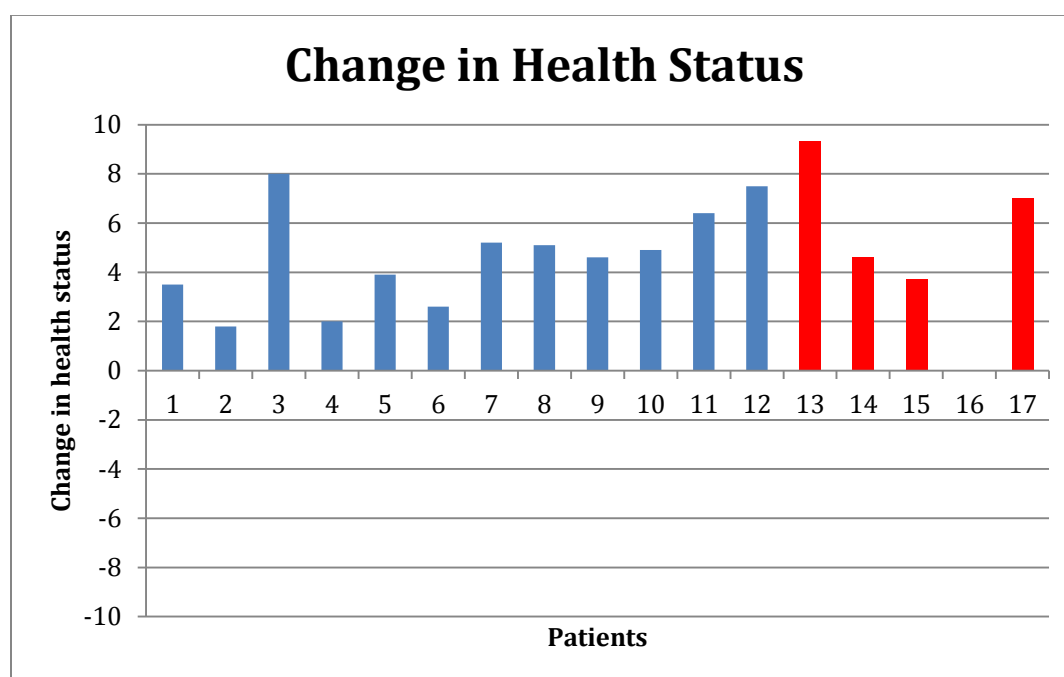


Figure 2. Change in health status

Change in health status

The visual analogue scale for the change in health status showed that all but one patient reported a positive improvement in health status after their BAHD for UCHL. One child reported no change. See figure 2.

Device usage

When looking at the number of days the BAHD was used by the children, in both the congenital and acquired groups, the majority were using their device 5-6 days a week.

When looking at the number of hours of usage a day, the majority of children were using the BAHD for at least four hours a day, in fact most of them were wearing it for more than eight hours a day. There was one child in the acquired CHL group that was using the BAHD for less than two hours a day.

Finally, 100% of children used their BAHD.

Satisfaction and Quality of life

None of the children were dissatisfied with the BAHD, in fact 16/17 (94%) were satisfied to a degree and only one child 1/17 (6%) felt that the BAHD made no difference. When questioned about how they perceived the BAHD had affected their quality of life, 13/17 (76%) children felt that it made a significant improvement. See figure 3.

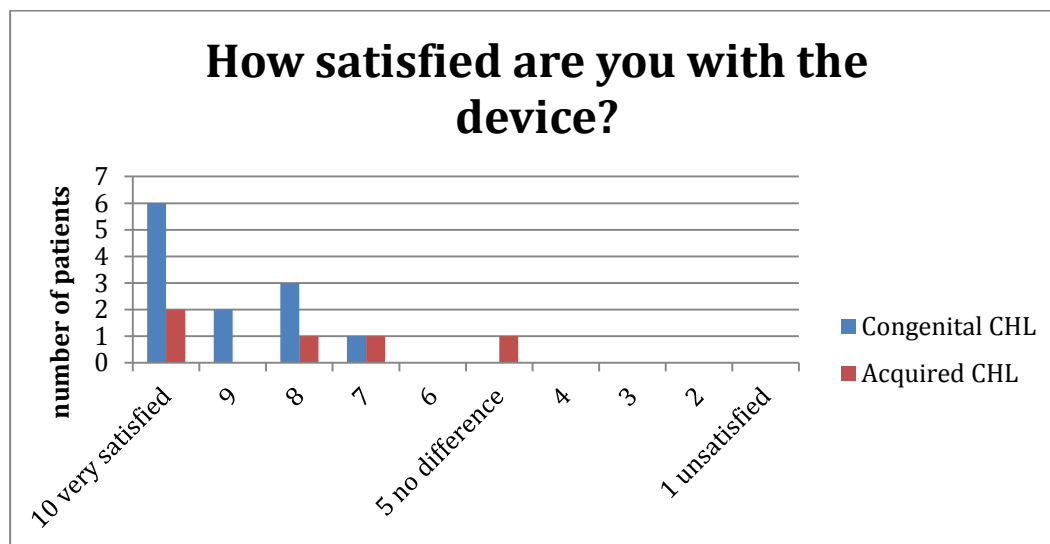


Figure 3. Satisfaction

The results from the SSD questionnaire reflected the value of BAHD in five specific situations: Talking to one person in a quiet situation, talking to one person in a group, listening to music, watching the television or radio and at the dinner table talking to a person on the deaf side. All of the children reported that their BAHD was of value in at least one of these conditions, with most of the children finding value in four or five of the specified situations. Looking at the situations in isolation, the BAHD was deemed most useful in a group situation and least useful talking to one person in a quiet background. The BAHD did not have a negative impact for any child in any of these specific situations. See Figures 4-6.

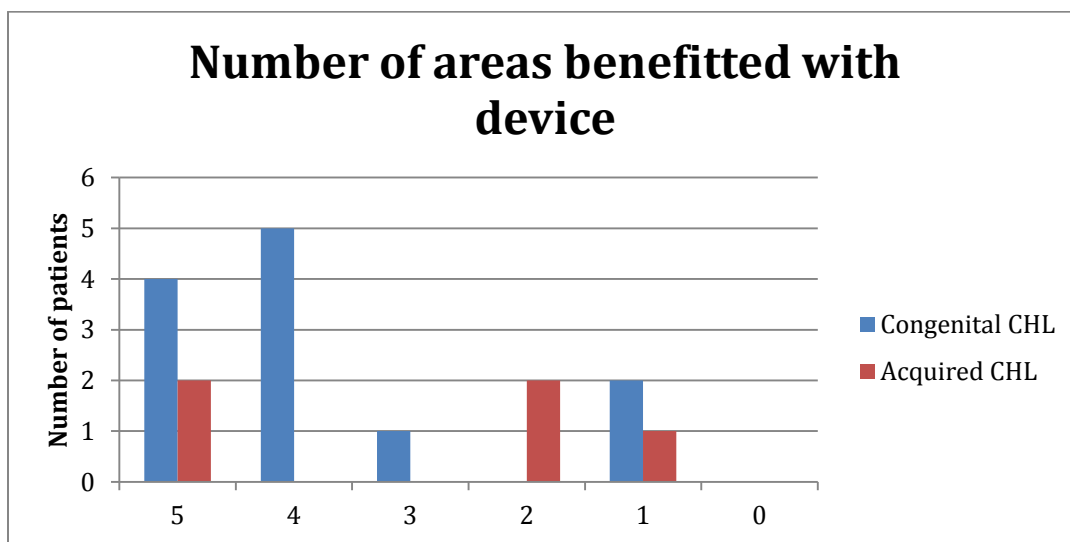


Figure 4. Number of areas benefitted with device

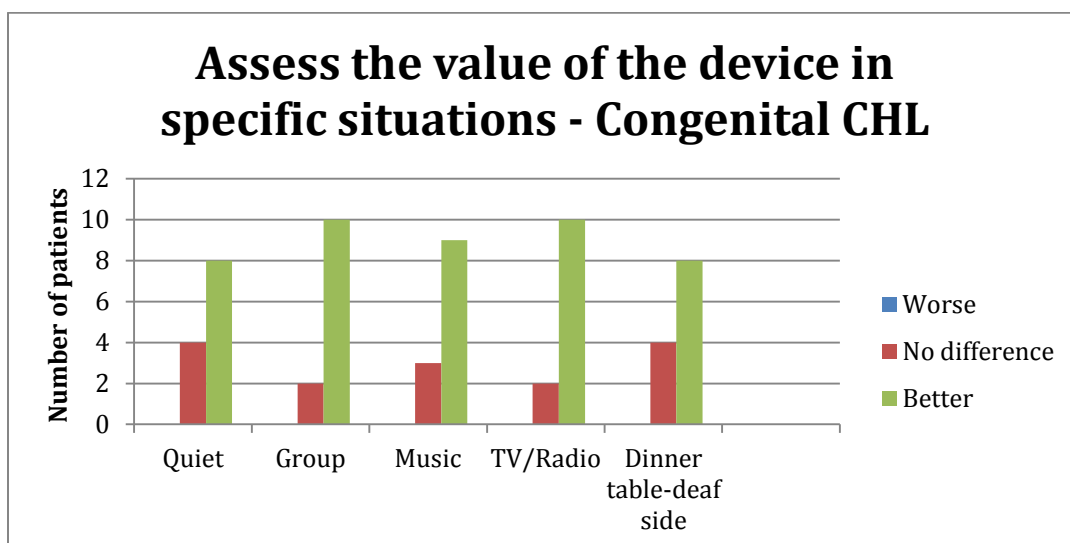


Figure 5. Value of device in specific situations - congenital CHL

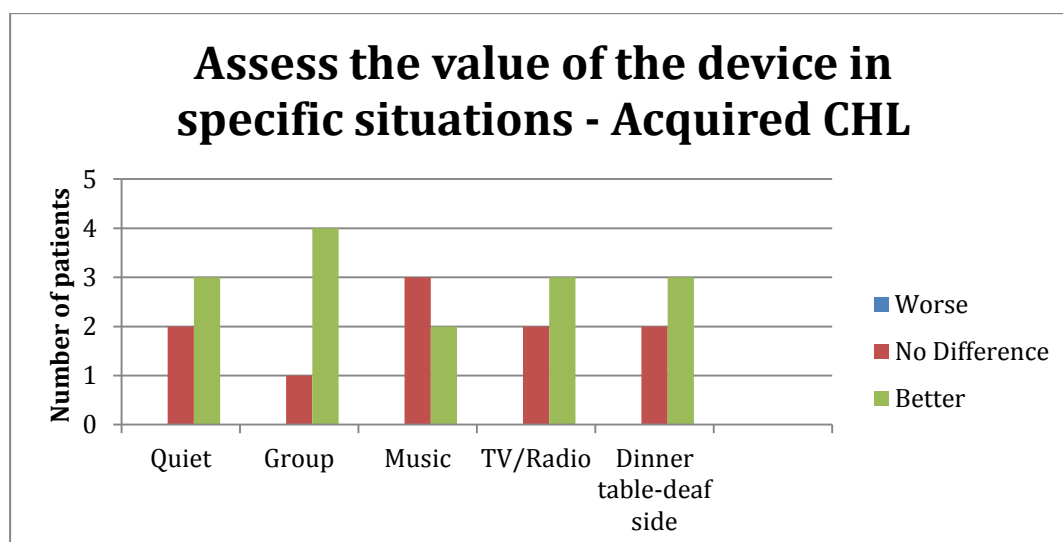


Figure 6. Value of device in specific situations – acquired CHL

Discussion

~~Any study using a patient questionnaire for results is subject to recall bias.~~

Carers are usually asked to help complete the questionnaires if the child is not old enough to do so themselves. It must be remembered that the results may reflect the carer's views and perceptions. When working with children the role of the parent/carers is hugely important and so their views and perceptions should be considered an important part of outcome results.

Evidence from studies using the GCBI have shown a prior expectation to the intervention and the perceived benefit from the intervention may deteriorate with time.¹⁵

The cohort of children in this study were 'a selected group' who had been referred for a specific hearing problem. All of the children were referred with significant speech and language difficulties, and/or significant educational and behavioural issues compared with their siblings or peers. The older children were noted to be having difficulties at school and were already having additional educational measures implemented.

Interestingly, 12 of the children had a clearly recognisable congenital UCHL with external ear abnormalities. The nature of their hearing loss was identified very early yet the age of referral for audiological assessment/ help was late (average age at referral being 7 years and 3 months).

All the children in this study had either a congenital or acquired CHL.

Only two children had additional learning difficulties (associated with co-existing medical conditions) that may have influenced their outcomes. When reviewing literature about children with any UHL, social backgrounds, medical co-morbidity and educational assessments all vary from cohort to cohort and so comparison of any of their outcomes is difficult. Another variable is the degree of support provided by different schools in different geographical areas. Some institutions cater for such children using many techniques including radio aids, individual class assistant's help and input from the visiting teachers of the deaf.

Further controversy surrounds the learning and behavioural problems. Have these arisen as a consequence of the long-standing hearing difficulties?

The educational potential of any child is dependant on many factors not just their hearing. Hence it is very difficult to interpret the actual benefit of the BAHD in any child with a UHL.

GCBI

The majority of the GCBI results were positive. One child had a negative score and two further children had small positive scores.

The child reporting a negative score was a teenager. This particular child had a number of obvious congenital abnormalities and had a long-standing tracheostomy in situ. Unusual chromosomal abnormalities had been identified but no formal syndrome or association had been diagnosed. Bullying had been a recurrent problem despite changing schools on a number of occasions. A significant self-image issue had resulted. Despite all of the above, this child reported an improved health status from their BAHD on the visual analogue scale. Furthermore, this child continues to wear their BAHD everyday.

Two children reported a small positive score on the GCBI: A teenage female with hemifacial microsomia who has concerns regarding her appearance and self-esteem. Again, despite her low self-esteem and issues with her image, she is a good user and reports hearing benefit with her BAHD. The second young man was also a teenager with similar issues regarding bullying and self-image although his UHL was acquired. Chronic ear discharge was the constant concern for him.

All three children scored poorly on the questions relating to emotion. They had issues with self-esteem and appearance. It would appear that for this group, the BAHD added to their negative self-image issues. Despite finding the BAHD of benefit, they were concerned about the appearance of the BAHD. This is a common problem in teenagers and adolescents. There is evidence in the literature that children, (particularly boys) have issues regarding self image when it comes to

bone conduction aids and BAHD¹⁸ which may adversely affect their questionnaire results.

The visual analogue results were interesting. Three of the children showed a small positive change on the GCBI. When compared to the corresponding visual analogue score, these three children showed a very large increase in health status. There was some subjective evidence from comments made in the free text that the BAHD had made a positive difference.

In the current health climate evidence supporting BAHD is crucial. Demonstration of positive self reported patient benefit resulting from a BAHD will be increasingly more important in the evaluation of a cost benefit analysis.

In the paediatric literature, de Wolf¹⁹ et al found that in children with congenital unilateral conductive hearing loss, the BAHD was of particular benefit in educational settings, but does not reliably lead to a significant benefit in all domains. It is re-iterated that in these children it is vital to perform a pre-operative trial with a headband in order to predict benefit.

In the adult literature Snik et al found that those patients with a longstanding or congenital hearing loss reported a smaller benefit than those with an acquired hearing loss.²⁰ Martin et al found that the BAHD was less beneficial in adults that had hearing loss for more than ten years.²¹

Not all children with a UHL need aiding, yet those with a significant hearing handicap do well with a BAHD. It is often very difficult for the BAHD clinician to decide what and when any treatment should be offered. Historically children presenting with a UHL were reassured if normal hearing thresholds were demonstrated in the contralateral ear.

Failure to identify those children with difficulties may likely result in a proportion of children who will not realise their full educational potential and be a burden to society.

In our institution, children referred with a UCHL are assessed audilogically with directional age appropriate hearing tests. They are fitted with a Baha® softband™ for a trial period of up to 3 months. They are advised to wear it both at school and in the home. At the child's school, the visiting peripatetic teacher is asked to report on their progress during this period along with other staff involved. In conjunction with this, the children or their carers fill in subjective quality of life questionnaires regarding their experiences with the Baha® softband™.

A BAHD is then offered if the trial period shows a significant improvement.

Carers/ parents of children presenting with a congenital CHL and congenital ear malformation are counselled regarding the possible long-term sequelae of unilateral hearing loss. Regular audiological assessment is arranged at age 9-12 months, 18 months, and yearly thereafter. Early intervention with a Baha® softband™ is offered if there are any concerns regarding hearing or speech development. Genetic counselling and discussion regarding cosmetic appearance of any congenital deformity of the ear are other aspects of the consultation.

Conclusions

The BAHD has an important role in the overall management of children with UCHL who are struggling with speech and language skills and have behavioural and educational issues. This study has shown a significantly improved quality of life in our cohort of children with symptomatic UCHL after receiving their BAHD. There was a high patient satisfaction and improvement in health status reported by children and carers. Qualitative subjective outcome measures demonstrated significant benefit. The vast majority of children had improved social and physical functioning as a result of better hearing and both carers and children reported an improved quality of life. Increased awareness of the potential consequences of a UCHL should be highlighted to healthcare professionals. An early opinion should be sought for any such child with difficulties that fail to respond to usual treatments.

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Appendix 1. The GCBI Questionnaire

1	Has your child's BAHA made his/her life overall better or worse?
2	Has your child's BAHA affected the things he/she does?
3	Has your child's BAHA made his/her behaviour better or worse?
4	Has your child's BAHA affected his/her progress or development?
5	Has your child's BAHA affected how lively he/she is during the day?
6	Has your child's BAHA affected how well he/she sleeps at night?
7	Has your child's BAHA affected his/her enjoyment of food?
8	Has your child's BAHA affected how self-conscious he/she is with others?
9	Has your child's BAHA affected how well he/she gets on with the rest of the family?
10	Has your child's BAHA affected his/her ability to spend time and have fun with friends?
11	Has your child's BAHA affected how embarrassed he/she is with other people?
12	Has your child's BAHA affected how easily distracted he/she has been?
13	Has your child's BAHA affected his/her learning?
14	Has your child's BAHA affected the amount of time he/she has had to be off nursery, playgroup or school?
15	Has your child's BAHA affected his/her ability to concentrate?
16	Has your child's BAHA affected how frustrated and irritable he/she is?
17	Has your child's BAHA affected how he/she feels about him/herself?
18	Has your child's BAHA affected how happy and content he/she is?
19	Has your child's BAHA affected his/her confidence?
20	Has your child's BAHA affected his/her ability to care for him/herself as well as you think they should, such as washing, dressing and using the toilet?
21	Has your child's BAHA affected his/her ability to enjoy leisure activities such as swimming and sports and general play?
22	Has your child's BAHA affected how often he/she needs to visit a doctor?
23	Has your child's BAHA affected how prone he/she is to catch colds or infection?
24	Has your child's BAHA affected how much medication he/she needed to take?

Appendix 2. The SSD Questionnaire

Question Number	Question	Possible responses
1	How many days/week do you use your device?	7days/week
		5-6 days/week
		3-4 days/week
2	How many hours/day do you use your device?	More than 8
		Between 4 and 8
3	Has your quality of life improved due to the device?	Yes
		No
4	Try to determine your satisfaction....(10 point rating scale)	Score from 0-10
5.1	Talking to one person in a quiet situation?	Better
		No difference
		Worse
5.2	Talking to one person among a group?	Better
		No difference
		Worse
5.3	Listening to music?	Better
		No difference
		Worse
5.4	Listening to TV/radio?	Better
		No difference
		Worse
5.5	At a dinner table, talking to a person sitting on your deaf side?	Better
		No difference
		Worse

Chapter 5

Peri-operative considerations for children undergoing Bone Anchored hearing Aid Surgery.

R. Banga, A. Reid, D. Proops, A-L. McDermott, M. Stokes

Abstract

Objectives: To identify important factors in the peri-operative management of children undergoing Bone anchored hearing device (BAHD) surgery in a paediatric tertiary centre. We also aim to compare current practice and identify any changes in practice with the previous study carried out in the same paediatric tertiary centre in 2000.

Methods: Children undergoing BAHD surgery between January 2008 and January 2011 were identified on a departmental database. A retrospective case note review was performed and compared with data collected prior to 2000.

Results: In the study period, 194 children were identified to have had BAHD surgery. 134 case notes were available for analysis and of these children, 353 anaesthetics were identified. 45.5% of the children had a recognised syndrome or dysmorphism and 17% had a congenital cardiac anomaly. 16% of the children were classified as a grade 3 or 4 laryngoscopy but 83.3% were managed with a laryngeal mask.

11.9% of the children had an intraoperative complication and 4.8% a postoperative complication. 88.4% of children were managed as day cases.

Compared with the previous study in 2000, there was a smaller proportion of syndromic or dysmorphic children and a larger proportion of children were managed with a laryngeal mask.

Conclusions: As BAHD surgery has become more common and as its indications have expanded, the perioperative management has evolved. The proportion of children with congenital heart disease has remained constant, but there has been a marked reduction in the number of children with syndromes involving the head and neck. We have found that even in complex craniofacial cases, the laryngeal mask is increasingly being used with good results. However, advanced paediatric airway experience was still required in a small number of cases, heightening the awareness that specialised paediatric support services are necessary for a comprehensive BAHD programme.

Introduction

The indications for Bone Anchored Hearing Device (BAHD) surgery have expanded since it was first introduced in the 1980s. In the adult population, the majority of BAHD surgery is performed as a single stage procedure, often under local anaesthesia. Children differ in that almost all their surgical intervention requires general anaesthesia and BAHD surgery in particular may require several anaesthetics. The main reasons are firstly, in many centres paediatric BAHD surgery is undertaken as a staged procedure. Secondly, the incidence of fixture failure and skin reactions in children is higher than in adults therefore the requirement for further surgical intervention is greater.¹ Lastly, there have been changes in BAHD funding in the National Health Service (NHS) in recent years and now children with bilateral hearing loss are commonly offered bilateral surgery.

Children suitable for a BAHD are a heterogeneous group and range from those with an isolated pinna malformation to the complex child with a significant congenital abnormality, syndrome and/or other co-morbidity. Surprisingly there is very little guidance in the literature on the management of these children in the peri-operative setting. In 2000, our colleagues at Birmingham Children's Hospital (BCH) reported the anaesthetic implications of BAHD surgery based on seven years' experience at that time; a cohort of 43 patients which required 102 anaesthetics.² The purpose of this study is to identify important factors in the peri-operative management of children undergoing BAHD surgery in a paediatric tertiary centre, placing particular emphasis on co-morbidity and the implications of airway management and anaesthetic technique. We also aim to compare current practice and identify any changes in practice with the previous study carried out in the same paediatric tertiary centre in 2000.

Methods

Children undergoing BAHD surgery between January 2008 and January 2011 were identified on a departmental database. A retrospective case note review was performed and information regarding all BAHD surgery was collected. At Birmingham Children's Hospital, children under the age of 10 years commonly undergo a two-stage procedure for a BAHD. At the first stage, two fixtures are implanted and typically, after a period of three months, the second stage is performed which includes a skin reduction and abutment fixation. Children over

the age of 10, undergo a single stage or, in some cases, two-stage procedure. Those children who had their initial surgery prior to 2008 were identified when they attended for either a second side implant or further surgery relating to their BAHD.

Data collected included patient demographics and co-existing morbidity. Details of the anaesthetic management recorded for each procedure included the method of induction of general anaesthesia. The view at direct laryngoscopy using Cormack and Lehane's classification³ was noted. In a grade I laryngoscopy view the complete glottis is visible, in grade II the anterior glottis is not seen, in grade III the epiglottis seen, but not the glottis, and finally in a grade IV laryngoscopy view the epiglottis not seen. Since the ease of view can change over time, we retrieved the laryngoscopic view for each anaesthetic.

Other data collected included method of airway management plus any abnormality necessitating special intervention. Both intra-operative and post-operative complications were noted.

A complication was defined as an event that was significant enough to require medical intervention.

Results

Patient demographics

We identified 194 children who underwent BAHD surgery between January 2008 and January 2011.

We retrieved the records of 134 patients. 60 patient records, however, were not available for analysis.

In the 134 children analysed, records of 353 general anaesthetics were identified. These related to first and second BAHD surgery, skin reductions and replacement fixture insertions. The number of procedures on each child ranged from 1 to 8 with an average of 2.6 operations per child. The age at time of surgery ranged from 2 years to 17 years, with both the mean and median age at surgery being 9 years and 1 month.

Co-morbidities

Syndromes and dysmorphism

Recognised syndromes and dysmorphisms were apparent in a significant proportion 61/134 (45.5%) of children. The most common were:

- Treacher Collins syndrome 15/61 (24.6%)

- Goldenhar syndrome 8/61 (13.1%)

Other dysmorphisms included:

- Pierre Robin sequence
- Chromosome 22q11 deletion
- CHARGE association
- Hemifacial microsomia
- Pfeiffer syndrome
- Foetal alcohol syndrome
- Branchio-oculo-facial syndrome
- Apert's syndrome.

The most common chromosomal abnormality was Down syndrome identified in 13/61 (21.3%) children. There was one child with trisomy 17, one with chromosome 7 deletion and two children with 18q deletions. The remaining children were otherwise healthy and were having BAHD surgery for either an isolated microtia / atresia or chronic suppurative otitis media.

Congenital Cardiac Anomalies

Cardiac problems were identified in 23/134 (17%) of children.

Of the 13 children with Down syndrome, 9 had structural cardiac abnormalities, all being defects of the atrio-ventricular septum. Eight (8/9) children had undergone corrective cardiac surgery before their BAHD procedure. The remaining child (1/9) was asymptomatic.

A further 9 children were noted to have other syndromes and cardiac abnormalities. All 9 had prior cardiac surgical palliation. The most complex child had Goldenhar syndrome and had undergone a Fontan palliation for tricuspid atresia.

5 children had structural cardiac abnormalities in the absence of a recognised syndrome or other abnormality. 3/5 of these children had minor atrio-ventricular defects and were systemically well. The remaining 2/5 had dysmorphic features and a more significant cardiac defect (tetralogy of Fallot and total anomalous pulmonary venous drainage, respectively).

Laryngoscopy grade and airway management

Laryngoscopy view grade 3 or 4 (implying difficulty in visualising the laryngeal inlet and vocal cords) was recorded in 57/353 (16%) cases. A grade 1 or 2 view (implying no such difficulty) was identified in 273/353 (77.3%). The laryngoscopy grade was unrecorded in 23/353 (6.5%) cases.

The most common method of managing the airway was by laryngeal mask (LMA) (83.3%), the remainder by endotracheal intubation. However, one LMA had to be replaced during the course of surgery and 6 required conversion to an endotracheal tube (ETT) intra operatively (Figure 1).

Cases recorded with a laryngoscopy grade 3 and 4:

Further analysis of this group revealed 24 children of which 19/24 had a recognised syndrome or malformation. These 24 children underwent 57 general anaesthetics for BAHD procedure. 41/57 (71.9%) were managed with a LMA and 16/57 (28.1%) were managed with an ETT.

Cases recorded with a laryngoscopy grade 1 and 2:

110 patients were identified in this group, of which 42/110 had a recognised syndrome or malformation. 273 procedures BAHD were performed for this group. 235/273 (86.1%) were managed with a LMA, and 34/273 (12.5%) were managed with an ETT and 4/273 (1.5%) were managed with an existing tracheostomy.

Induction of Anaesthesia

Intravenous induction of anaesthesia was chosen in 199/353 (56.4%) cases, most commonly with propofol (Diprivan marketed by AstraZeneca) (182/199). A small number of patients (17/199) received sodium thiopentone. Where induction was inhalational 149/353 (42.2%), the vapour was invariably sevoflurane. The induction agent was not recorded in 3/353 (1.4%) cases.

Complications

Intra operative complications

The intraoperative course was uneventful for 87% of BAHD procedures. (308/353) However complications occurred in 42/353 (11.9%). No information was available in 3/353 (0.8%) cases.

Most problems encountered were airway related, predominantly due to malpositioning or dislodgement of the LMA. In six patients the LMA had to be changed to an ETT intraoperatively. In one patient the problem was resolved by replacement LMA.

One otherwise fit and healthy child with no co-existing co-morbidities had peri-operative aspiration and bronchospasm.

Fibreoptic intubations were recorded for two children in this series. Both children had severe retrognathia. One had a cleft palate and the other had significant difficulties with delayed bone maturation, skeletal immaturity and poor growth.

Binder syndrome was found to be associated with a very difficult intubation and required Paediatric Intensive Care. This child was found to have a grade 3 sub-glottic stenosis (>70%) on the Myer - Cotton grading system⁴ but had a background of “severe asthma.” A tracheostomy and later laryngotracheal reconstructive surgery were necessary prior to proceeding with the BAHD surgery.

Postoperative complications

Postoperative complications were encountered in 17/353 (4.8%) cases. The postoperative course was uneventful in 333/353 (94.3%) cases. No information was available in 3/353 (0.8%) cases.

The most common documented problem was postoperative nausea and vomiting, encountered on 6 occasions. Three further patients required oxygen overnight and one patient required treatment for bronchospasm.

Day case procedures

312/353 (88.4%) procedures were performed as a day case, 39/353 (11%) stayed in hospital overnight and this information was not documented in 2/353 (0.6%) cases.

Those children that suffered an intra operative airway complication appeared to recover well and return home the same day.

Those children that required an overnight stay had either a postoperative complication requiring oxygen and or a bronchodilator, an intraoperative complication with other medical co morbidities or resulted from unmanageable geographical difficulties and travel arrangements.

Discussion

Patient demographics

The marked increase in the number of children undergoing BAHD surgery at our institution in the current study period is most likely due to an increased awareness of BAHD amongst other healthcare professions, the expanding indications for BAHD and also the increased knowledge and demands of patients and their carers. Specifically in the earlier 7 year study period, 43 children were recorded to have undergone a total of 102 anaesthetics for BAHD surgery.² In our 3-year study period we identified 194 children that had had BAHD surgery. Of the 134 case notes analysed, there were 353 anaesthetics performed, with 135 of the anaesthetics falling within the study period.

The cohort of children in this study was also very different to the previous study. In particular, there was an increase in the proportion of otherwise medically fit children with acquired hearing loss, specifically unilateral hearing loss, and just 45.5% of this study group had an associated syndrome, malformation or other medical co-morbidity. Fewer children had craniofacial abnormalities. Although the percentage of children with Goldenhar's syndrome (13.1 versus 26%) was reduced, the number of children with Treacher Collins syndrome remained constant (21% versus 24.6%). Previous studies have shown that up to 59% of children on a BAHD programme can have significant co-morbidity of which craniofacial abnormalities are the most common.⁵

Changes in anaesthetic techniques and anaesthesia.

The approach to anaesthetic management has changed between the study periods reflecting both the changing cohort of patients and shorter duration of operations. This is most notable in the intraoperative management of the airway. Most children were managed with a LMA (83%). Since the surgical procedure has evolved and surgical time has reduced, the use of a LMA as a first choice has become more popular, even in some of the more complex children. However, problems did occur with the use of a LMA, mostly due to malpositioning and early conversion to ETT intubation was needed in a small number of cases.

The use of the LMA is controversial in children with known craniofacial abnormalities but they are increasingly being used with good results, even in those with known difficult airways including craniofacial abnormalities.⁶ Problems are few, but do occur and in our series advanced paediatric airway experience was still necessary in 7 children and one child required Paediatric Intensive Care. This should heighten awareness that specialised paediatric support services are necessary for a comprehensive paediatric BAHD programme.

The view on laryngoscopy in most children in this study was graded either 1 or 2 on laryngoscopy (77.3%). This is a higher proportion when compared to the previous study carried out in 2000 (67.4%). Similarly, there were fewer children graded 3 or 4 at laryngoscopy (16% versus 32.6%) further highlighting the increased number of medically uncomplicated children who can now be considered for a BAHD. It was very helpful to have this laryngoscopy view for previous procedures documented to plan airway management.

In this context of a consultant delivered service, the patients benefitted from experienced anaesthetists and surgeons who regularly manage complex cardiac and other paediatric airway procedures.

Interestingly, the choice of inhalational or intravenous induction technique was not noted to show any trend and was predominantly patient and family choice. The infrequent use of induction agents other than propofol was most likely a reflection of wider and current anaesthetic practice.

Although postoperative nausea and vomiting are recognised complications of general anaesthesia, the incidence in this group of children was less than 5%. None of the children received opiates. Simple analgesics such as paracetamol and non-steroidal anti-inflammatory medications were the analgesics of choice in combination with local anaesthetic infiltration of the surgical site. At the time of this study, it was not routine practice to administer anti-emetics in this group of children however current practice has changed and now it is more common. This approach has enabled the majority of children to have BAHD surgery as a day case procedure.

Increasing numbers of BAHD centres are reporting single stage BAHD surgery for children of any age.⁷ Although on first glance this does reduce the number of general anaesthetic required, a BAHD programme for children has other challenges which may incur the need for further surgery, including a higher risk of trauma, failure of osseointegration and also peri-abutment skin reactions. Our experience would suggest that multiple anaesthetics are generally well tolerated and do not preclude a staged approach to BAHD surgery.

Conclusions

In our experience, the factors that we would suggest are important are: Thorough pre-assessment, good record keeping and communication and an experienced surgical and anaesthetic team who are familiar with the anatomical and medical co-morbidities that may be present. The majority of children are suitable for day case procedures

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Chapter 6

Subjective benefit following the addition of a second BP100 softband processor for children with bilateral conductive hearing loss.

R. Banga, A. Child, A. Reid, A.-L. McDermott

Abstract

Introduction: It has been shown that children with bilateral hearing loss benefit with bilateral implantation with a bone anchored hearing device (BAHD) once old enough. Since very young children have a higher complication rate with a BAHD, it is common practice in many centres to trial a sound processor on a soft headband until the skull is thick enough for implantation. Currently, this is typically delivered as a unilateral processor on a headband.

The aim of this study is to evaluate whether a bilateral sound processor on the Baha softband® will confer additional subjective benefit.

Methodology: 14 children were identified with bilateral conductive hearing losses. They all underwent a trial of a softband® with one BP100 sound processor for a minimum of 3 months after which time all children were then fitted with a second BP100 processor i.e. a bilateral Baha softband®. Each child was then evaluated after a further 3 then 6 month period of bilateral Baha softband® use.

Questionnaires were used at each visit.

Preschool children's carers completed both the Glasgow Children's Benefit Inventory (GCBI) questionnaire and the Infant-Toddler Meaningful Auditory Integration Scale ITMAIS questionnaire.

School age children and their carers completed the GCBI and the Children's Home Inventory for Listening Difficulties (CHILD) questionnaires.

Results: All of the children demonstrated a subjective, significant and sustained improvement with the addition of a second BP100 processor.

Conclusions: All children with bilateral hearing loss should be fitted with bilateral sound processors on a headband – particularly in the under 5-age group.

Introduction:

Children with hearing impairment that are identified and rehabilitated before the age of six months develop significantly better language scores than those identified at a later stage.^{1,2} But it is also well documented in the literature that young children have a higher complication rate following BAHD surgery when compared with adults.³⁻⁶

Children with hearing loss fitted with sound processors on soft and hard headbands have been shown to develop good speech and language skills and manage educationally on par with their peer groups. In 2002 the Baha softband® was introduced as an alternative to the bone conductor on a steel headband. It has been found that the Baha Softband® provides better audiological outcomes when compared with traditional bone conduction aids.^{7,8} Since the results with the Baha Softband® have been so good, the need for surgical intervention in younger children with higher has been eliminated.⁶ Clinicians now have the confidence to wait until the child is of sufficient age and size to allow for successful implantation without a negative impact on the child.

Traditionally children with a bilateral conductive hearing loss have been fitted with a unilateral sound processor headband. The rationale being, that one bone conductor will facilitate hearing in both cochleae due to the minimal attenuation of sound waves across the skull.^{9,10}

Previous studies on adults have demonstrated that bilateral bone conductors provide the patient with improved sound localisation, speech recognition in quiet and to some extent, speech recognition in noise.¹¹⁻¹³

Dun et al looked at quality of life outcomes in 27 children and young adults fitted with bilateral BAHDs for bilateral conductive hearing losses. They found that in the vast majority bilateral BAHDs provided better hearing quality, with better spatial hearing in those fitted binaurally at a young age.¹⁴ Audiological testing in a similar cohort of patients showed that directional hearing is improved with binaural BAHD fitting however it is not clear which auditory clues effect this advantage.¹⁵

Aim: To establish whether there is any additional subjective benefit for a child with a bilateral conductive hearing loss currently using a unilateral Baha Softband® to have the addition of a second sound processor.

Methodology

14 children were identified with a bilateral conductive hearing loss. All children were currently using a Baha Softband® with a unilateral BP100 sound processor on the poorer hearing side for more than a 3 month period.

All carers, and children when age appropriate, completed validated questionnaires before the fitting of a bilateral BP100 Baha Softband®. These validated questionnaires were repeated following both a three month and six month period of bilateral Baha Softband® use.

Questionnaires used in the study

Pre school children: (IT-MAIS) Infant-Toddler Meaningful Auditory Integration Scale (see appendix 1), (GCBI) Glasgow Children's Benefit Inventory (see appendix 2), Health Status.

School age children: (CHILD) Children's Home Inventory for Listening Difficulties (see appendix 3), Glasgow Children's Benefit Inventory (GCBI), Health Status.

The IT-MAIS is a modification of the Meaningful Auditory Integration Scale (MAIS) developed by Advanced Bionics.¹⁶ It is a structured interview designed to assess a child's responses to sound in everyday environment. The parents/carers are interviewed in three main areas: vocalization behaviour, alerting to sounds and deriving meaning from sound. The scoring is calculated based on each question (0-4 points) and the maximum score is 40.

The CHILD questionnaire designed by Oticon¹⁷ is a 15-item questionnaire for the parent and child to fill in regarding typical family communication situations at different distances and in background noise. Each question scores a value of 1-8 and an average score is calculated for the parental and the child filled questionnaire.

The GCBI is a validated subjective child orientated post-interventional questionnaire designed to evaluate any paediatric otolaryngology intervention.¹⁸ It consists of 24 questions based upon a five point Likert scale. A score of +2 shows a maximal positive change and a score of -2 a maximal negative change. The sum total score is divided by 24 and multiplied by 50 to give a score ranging from +100 to -100 depicting a positive or negative change. Specific questions in the inventory relate to emotion, physical health, learning and vitality.

We additionally asked the children/carers to indicate their perceived health status before and after intervention on a linear visual analogue scale. This has been used in other published quality of life papers and has been shown to correlate with GCBI score.¹⁹⁻²²

Results

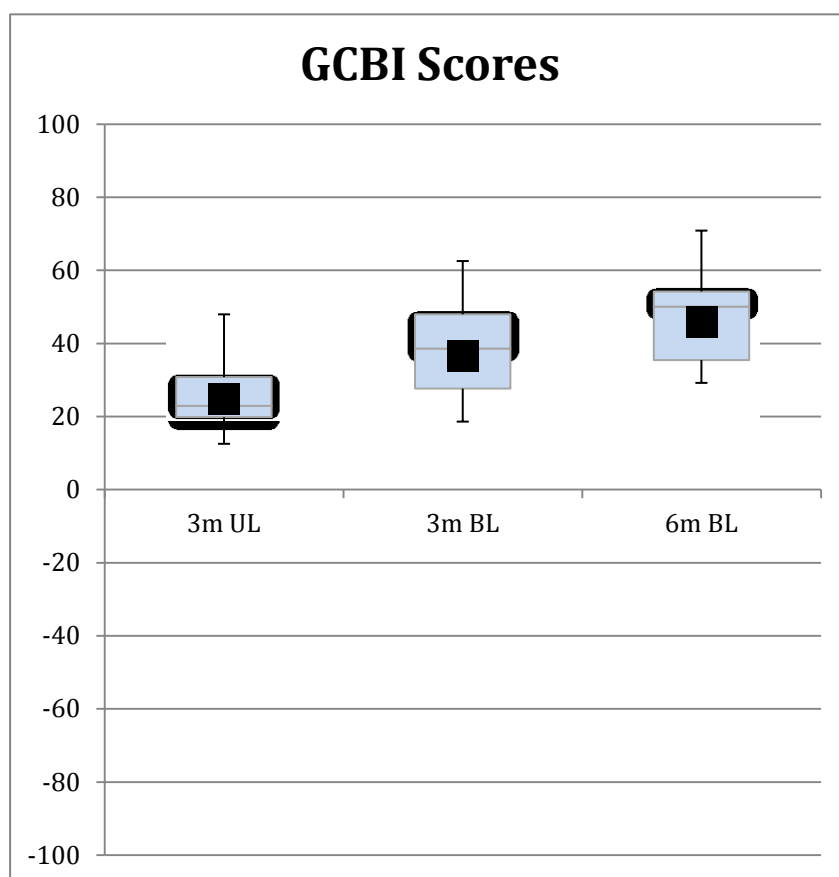
14 children were identified with a bilateral conductive hearing loss that had been wearing a Baha Softband® for a minimum of 3 months. There were 6 girls and 8 boys with ages ranging from 2 years to 12 years.

Pt	Age	Gender	Medical History	Previous hearing rehabilitation
(1)	3	F	Down syndrome, Tracheostomy, tiny ear canals	Unable to tolerate conventional AC aids. Non compliant with conventional BC aid
(2)	3	M	Goldenhar, Bilateral atresia	Unilateral compact BAHA processor on softband since 5/12 of age
(3)	3	M	Nagers Syndrome, Bilateral atresia	Unilateral compact BAHA processor on soft sports band since age of 2/12
(4)	2	M	Chromosomal Abnormalities, Bilateral atresia	Unilateral conventional BC aid attached to a soft sports band for the last 4 months
(5)	3	M	Treacher Collins, cleft palate	Bilateral conventional AC aids
(6)	2	M	Treacher Collins, bilateral microtia and atresia	Unilateral compact BAHA processor on a softband for 4 months
(7)	3	M	Down syndrome, tiny ear canals	Unilateral Conventional AC aid
(8)	2	M	Down syndrome, tiny ear canals	Unilateral BP100 processor on a softband for 18/12
(9)	4	M	Falconi anaemia, lung lobectomy, bilateral atresia and microtia	Unilateral compact BAHA processor on softband since 7/12 of age
(10)	2	F	Treacher Collins with tracheostomy and speaking valve	Unilateral compact BAHA processor on softband since 7/12 of age
(11)	3	F	CHARGE	Unilateral conventional BC aid for a year
(12)	6	F	Treacher Collins	Unilateral compact BAHA processor on a softband ?how long
(13)	12	F	CSOM, multiple mastoid surgery aged 9/10	Bilateral conventional AC aids ?how long
(14)	9	F	BL canal stenosis - attempted surgical correction failed	No previous aiding

Pt	Average BC Thresholds (dB HL)	Average Free Field AC Thresholds (dB HL)	Average Left AC Thresholds (dB HL)	Average Right AC Thresholds (dB HL)
(1)	13	50	-	-
(2)	10	65	-	-
(3)	7.5	-	57.5	60
(4)	16.3	61.7	-	-
(5)	-2.5	-	27.5	47.5
(6)	5	-	60	65
(7)	15	61.3	-	-
(8)	10	58.8	-	-
(9)	15	-	58.8	70
(10)	5	46.7	-	-
(11)	11.7	-	38.8	55
(12)	5	-	61.3	57.5
(13)	-1.25	-	58.8	65
(14)	0	-	41.3	47.5

In the group with 11 younger children (patient numbers 1-11), 2 children (patient numbers 4 and 11) were implanted prior to the end of the study period as they benefitted so much from the addition of the second processor and did not wish to delay implantation. Hence in this group all 11 carers were able to complete GCBI and IT-MAIS questionnaires at 3m UL and 3m BL stages. The remaining 9 carers completed the questionnaires at 6m BL.

Only 3 children (patient numbers 12-14) were emotionally mature enough to complete the CHILD questionnaire, however, in this group, none of the children have worn the second processor for more than 3 months, so questionnaires are only available for the 3m UL and 3m BL stage.



3m UL - Questionnaires completed after 3months in a unilateral sound processor
 3m BL- Questionnaires completed after 3months in a bilateral sound processor
 6m BL- Questionnaires completed after 6months in a bilateral sound processor

Figure 1. GCBI scores – Box and Whisker Plot

After 3 months with a unilateral processor the average GCBI score was 26. All 14 children displayed an improved GCBI score 3 months after the addition of a second processor on the softband, with an average score of 38. The GCBI score improved further at the 6-month stage to an average of 47.

We also asked all children/carers to report the change in health status on a linear visual analogue scale before and after interventions. Prior to the study the average reported health status was 2.8. After 3 months with a unilateral processor the average reported health status was 7.2. 3 months after the addition of a second processor the average reported health status was 8 and after 6 months it was 8.4. 11 out of the 14 children were not emotionally mature enough to fill in questionnaires and thus their carers were asked to fill in the IT-MAIS Questionnaire. 3 months after the addition of the second processor, the average IT-MAIS score increased from 25 to 31.

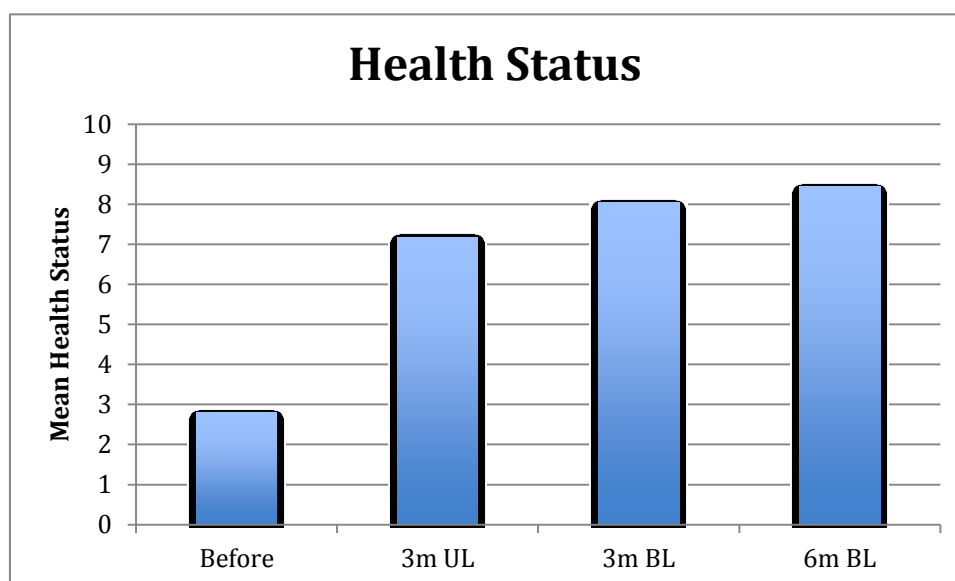


Figure 2. Health Status

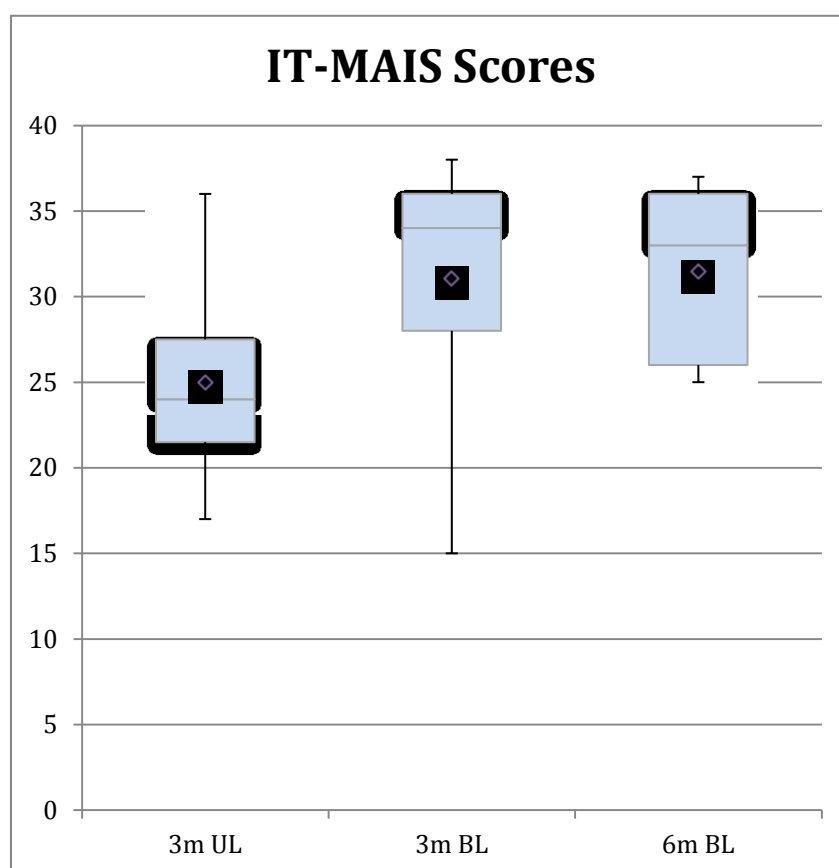


Figure 3. IT-MAIS Box and whisker plot

This benefit was sustained at 31 at the 6-month stage. There were a small group of children who initially worsened after the addition of the second processor, evident from the lower whisker. The most common scenarios that they found difficult were responding to their name with no visual cues and differentiating between speech and non-speech stimuli. In all cases, at the 6m BL stage they had overcome this difficulty. The majority of these children had been using monaural rehabilitation prior to the study and we feel that this short lived worsening in the IT-MAIS scores may reflect an adaptation period necessary to adjust to binaural hearing after a period of monaural hearing.

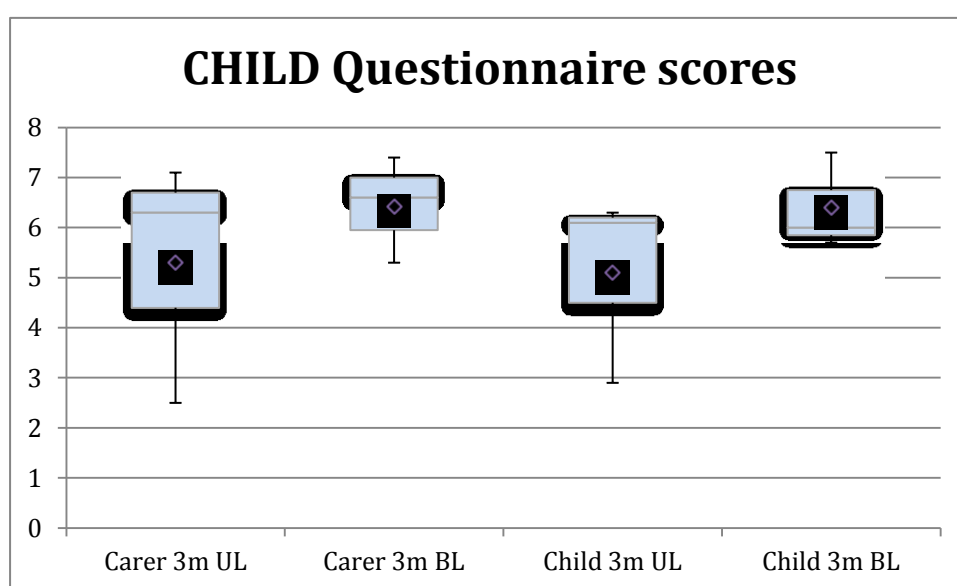


Figure 4. CHILD – Box and whisker plot

The three children that were emotionally mature enough to fill in a questionnaire were asked to complete the CHILD Questionnaire and their carers also completed the adult version of the CHILD Questionnaire.

After 3 months with a unilateral processor, the mean CHILD score was 5.3 for the carers and 5.1 for the children. There was a further improvement in scores with the addition of a second processor with both carers and children scoring 6.4. None of these children have been wearing the bilateral processors long enough to complete the final questionnaires as yet.

A subjective improvement was identified in all children following three months using bilateral BP100 processors on a softband. Further assessment at six months

demonstrated a subjective and sustained improvement with the addition of the second BP100 processor in all the children irrespective of their other co-morbidities.

Discussion

There is sufficient evidence that binaural hearing is better than monaural hearing resulting in improved subjective and objective outcome measures^{13,23} and that spatial hearing is correlated with bilateral fitting at a younger age.¹⁴ In addition, it has been shown that in longer-term studies, adults with bilateral hearing losses that are aided binaurally achieve higher secondary school qualifications when compared with those that had been aided monaurally.²⁴

With the advent of universal screening programmes, children with bilateral hearing losses are being detected at a younger age, when they may not be suitable for surgical intervention. The advent of soft and hard headbands have enabled surgery to be delayed until adequate skull maturation is achieved, but this has resulted in children spending their formative years typically being aided monaurally in a headband with only one sound processor. The literature suggests that bilateral aiding is superior to unilateral aiding in those with bilateral hearing losses. Some centres are implanting children with bilateral hearing losses with bilateral BAHDs after spending their formative period in a unilateral headband; thus it should follow that whilst they are in their formative years awaiting skull maturation that they should also be aided binaurally on a headband.

In some children there may be a period of “adaptation” if they had only monaural aiding previously. Therefore it is important to persevere with a suitable length of bilateral sound processor headband trial. The authors would recommend fitting of bilateral bone conductor headbands at their first presentation to the bone anchored hearing clinic.

Conclusions

All children with bilateral conductive hearing loss should be fitted with a bilateral bone conduction headband prior to any bone anchored hearing device implantation.

The practice of using a unilateral bone conduction device in these children should be discouraged.

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Chapter 7

General discussion

The past decade has seen the introduction of numerous implantable hearing rehabilitation devices. The percutaneous BAHDs have been evaluated extensively and the evidence in the literature tells us that they are effective and safe. The next few years will be very exciting in implantation otology with the advent of newer transcutaneous and ultimately fully implantable options, which will need the same rigorous scientific evaluation to provide patients and clinicians with the best treatment options.

Within the National Health Service in the UK, financial considerations are important in order to provide the highest cost-benefit treatments. Increasingly patient reported outcome measures such as quality of life outcomes are being used as tools to evaluate treatments, particularly in the paediatric setting where quantitative measures such as audiological testing are more challenging to achieve.

Chapter 2 Bone Anchored Hearing Devices in Very Young Children

This chapter evaluated the outcomes of BAHD in children aged 5 years and younger. There has been some controversy regarding implantation in this young age group in the literature. There are higher reported morbidity and fixture failure rates when compared with older children and adults. The findings revealed that in this young age group, the morbidity was higher. However, with the introduction of the softband indications for implantation in this young age group have diminished.

Chapter 3 Quality of life Outcomes following Bone Anchored Hearing Device Surgery in Children with Single Sided Sensorineural Deafness.

This chapter evaluated the BAHD used in children with a unilateral sensorineural hearing loss. The adult literature has suggested that there are mixed outcomes in implantation for single sided deafness. Four children with a congenital hearing loss and four with acquired loss were implanted. All but one child displayed an improvement in GCBI scores. There was a high degree of patient reported satisfaction.

There is very little evidence in the literature regarding the use of BAHDs for single sided sensorineural deafness in children.

Both this paper from Birmingham Children's Hospital and a further paper from the Arkansas Children's Hospital³ are at the time of writing the only papers that evaluate the single sided deafness application in children. These are small series and in time this application needs to be evaluated in larger numbers, perhaps combining data from several centres to gain more power.

Both studies found an improvement in patient reported subjective outcome measures and quality of life, often out of proportion to the measurable audiological benefit. It seems that the trial of headband is one of the most important factors in predicting the likely benefit and future studies must be performed in order to show whether there are improvements in long term educational goals rather than using audiology as the main outcome measure.

Chapter 4 Bone Anchored Hearing Aids in Children with Unilateral Conductive Hearing Loss. A patient/carer perspective.

There is some debate as to the necessity of aiding children with unilateral hearing loss as some children cope very well and others struggle with educational demands. This chapter looked at the quality of life outcomes in children with symptomatic unilateral conductive hearing loss that were implanted with BAHD. 17 children were evaluated and a high degree of patient satisfaction and improvement in health status was found after implantation. Research from Nijmegen has shown that individuals with unilateral conductive hearing losses show a varied performance on audiological testing¹ and that patients with an acquired unilateral conductive hearing loss performed better than the congenital group when evaluating sound localisation². This might be attributable to the fact that these children present for intervention only when there is already an existing problem with speech language or education. In the congenital group this means that there will have already been a prolonged period of time with out auditory stimulation in one ear, giving more strength to the hypothesis that bilateral auditory stimulation in the early years creates better performance in the long term.

Chapter 5 perioperative considerations for children undergoing Bone anchored hearing aid surgery

Chapter 5 looked in detail at the complexities of children on a BAHD programme at a tertiary referral centre. These children often require multiple surgeries and have a number of co-existing morbidities. 45% of children on the programme had a recognised syndrome or dysmorphism and 17% had a congenital cardiac anomaly. Despite the high number of potentially difficult airways, most children were managed with a laryngeal mask airway and were performed as a day case procedure. Senior airway management skills were still required in a few cases, even in those children with no risk factors.

This paper has shown that there is a significant proportion of children with complex medical needs. These children often require sequential imaging to monitor their conditions and in the paediatric population the modality of choice is magnetic

resonance imaging, as it does not involve radiation. The newer transcutaneous devices that require a magnet to be implanted may pose a problem in this cohort of children that require repeated imaging due to the movement and heating of the magnet and also due to the artefact created by the magnet.⁴ Further evaluation on the number of children that require imaging is needed and experiences from other paediatric and adult centres will be invaluable in order to best decide which device will be suitable for which child. There is an argument for centralising these services in dedicated paediatric centres.

Chapter 6 Subjective benefit following the addition of a second BP100 softband processor for children with bilateral conductive hearing loss.

This chapter evaluated the additional subjective benefit derived from a second sound processor in children with bilateral conductive hearing loss. All 14 children demonstrated improved and sustained quality of life outcome measures with bilateral sound processors when compared to a unilateral sound processor. This has implications for the auditory rehabilitation of young children prior to the time when they might be suitable for surgery.

This early bilateral stimulation of the auditory pathway may help to prevent children from developing a dominant and weaker central auditory pathway, if aided unilaterally. It has been suggested that children with a congenital unilateral conductive hearing loss that have not been aided at a young age find it difficult to achieve true binaural hearing for this reason.⁵ Further research will be needed to confirm these results with longer follow up and larger case series and perhaps additional audiological directional testing.

Future directions

The long-term benefits of auditory rehabilitation need to be investigated in larger groups. We need to look not only at quality of life outcome measures and audiological data, but also to look at the long-term educational outcomes of these children to determine what level of hearing loss requires aiding. How much difference does a 10 dB hearing loss make? It is a relatively recent discovery that children with unilateral hearing losses have poorer educational outcomes compared with their peers – so if we studied children with varying degrees of hearing losses i.e. 10, 15, 20 dB how much would this influence their final educational outcome?

This has some very important implications in the development of future devices. We are already beginning to see that the audiological output of transcutaneous devices such as the Sophono is not comparable with the percutaneous BAHD with

around a 10 dB difference.^{6,7} With the addition of background noise in the classroom setting, perhaps they are not good enough at present?

It remains to be seen if upcoming competing passive and active transcutaneous devices will overcome this problem.

Recent developments with different abutment sizes, shapes and surfaces have also increased the number of surgical options available to clinicians. These newer abutments may be more stable in bone due to a larger surface area of contact, but will this in turn cause an increased number of skin reactions, particularly in the paediatric setting? This is yet another area that will require thorough evaluation, particularly in light of the fact that newer abutment and fixture designs are not compatible between different manufacturing companies.

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Chapter 8

**Summary/Samenvatting
Curriculum Vitae
Acknowledgements
List of Publications**

Summary

Bone anchored hearing devices (BAHDs) have been successfully used commercially for the last three decades. Initially the indications were for those that were unable to wear conventional hearing aids. Typically these were patients with chronic middle and external ear disease or those with congenital malformations of the pinna or meatus. As clinical experience has increased and the rehabilitation results have been good and well accepted, the indications have expanded and now BAHDs are used in patients with unilateral and bilateral hearing losses both of a conductive and mixed nature. This thesis focuses on some of the more recent indications and special considerations in the paediatric population, in particular using patient reported quality of life outcome measures.

The lower age limits of implantation have been explored and in parallel to the literature this thesis finds that the fixture failure rate and morbidity in the under five group is higher than in older children and adults. Practice in this young group of children has since changed, especially with the introduction of the softband. This has enabled younger children to be adequately aided without the associated increased morbidity with BAHD in this younger cohort.

It is well known that bilateral hearing loss causes dramatic problems with speech and language acquisition, behaviour and also long term educational outcomes. As we obtain more experience with BAHDs and begin to understand the impact auditory rehabilitation has on the developing child, the importance of binaural hearing is being realised.

In adults with single sided sensorineural deafness, the BAHD has shown improved audiological and subjective outcome measures compared with the unaided state. The quality of life benefit may not be as marked when compared with other indications for BAHD. There is much less experience with this indication in children, however in this thesis we found that the carers reported a perceived improvement in health status and quality of life measures.

In children with a unilateral conductive hearing loss, the impact of their hearing loss can be very variable. Some children cope very well and others struggle. This thesis (and indeed most of the literature that evaluates such children) tends to focus on the children that are already experiencing morbidity from their hearing loss, as these are the families that seek audiological rehabilitation. These children

will already have had a prolonged period of unilateral auditory deprivation, and although carers reported improved quality of life measures, perhaps children would gain more benefit if aided prior to developing difficulties at school.

Increasingly, the literature shows that binaural hearing is superior to monaural hearing, and that binaurally aided patients achieve better long-term educational goals compared with those that are monaurally aided. Children with bilateral hearing losses are being detected at an early age and yet typically aided monaurally in a headband with only one sound processor in their formative years. This thesis demonstrates that the addition of a second sound processor results in improved and sustained quality of life measures, adding more evidence to the need for binaural aiding.

The BAHD programme at Birmingham Children's hospital has been running for twenty years and as it is a tertiary referral centre, some of the children have very complex medical needs and/or significant craniofacial anomalies. Most surgical interventions require general anaesthesia in children and many centres are performing staged surgery for BAHD in children. In addition to this, the BAHD complication rate requiring revision surgery in children is higher than in adults, hence the need for multiple anaesthetics. As the programme has developed and BAHD surgery becomes more common, there has been an increase in the proportion of medically fit children being implanted. Most are managed with laryngeal mask airway – even those with the complex craniofacial problems. Despite a low complication rate, in a small proportion of cases advanced paediatric airway skills were still required heightening the awareness that specialised paediatric support services are necessary for a comprehensive BAHD programme.

Samenvatting

De toepassing van het in het schedelbeen verankerde beengeleider hoortoestel (BAHD) geschiedt nu al weer zo een dertig jaar heel succesvol. Aanvankelijk werd de toepassing vooral beperkt tot die slechthorenden, die onoverkomelijke problemen hadden met de toenmalige conventionele luchtgeleidings en beengeleidings hoortoestellen. Dan ging het vooral om personen met een dubbelzijdige conductieve (gelegen in gehoorgang of middenoor) of gemengde (conductieve en binnenoor) slechthorendheid als uitkomst van een resttoestand van een chronische middenoorontsteking of een dubbelzijdige aangeboren gehoorgangatresie al of niet tevens met een microtie/anotie van de oorschelp.

Toen eenmaal gebleken was, dat de behandeling met de percutaan verankerde beengeleider hoortoestellen (BAHD) – zelfs tot de verrassing van velen - zo enorm succesvol was ook vanwege de zo goede revalidatie van het aanwezige gehoorverlies, werden het aantal behandelde patiënten en het aantal centra die deze behandeling gingen aanbieden al rap groter. Inmiddels zijn wereldwijd anno 2012 al meer dan 100.000 personen met een BAHD behandeling gerevalideerd.

De toepassing van de BAHD behandeling werd al snel uitgebreid van aanvankelijk een laatste mogelijkheid voor revalidatie van een problematische groep patiënten tot een voorkeursbehandeling vanwege grote eenzijdige en dubbelzijdige conductieve en gemengde gehoorverliezen. Al langere tijd is ook de behandeling van eenzijdige grote binnenoorverliezen bij volwassenen een behandeloptie geworden door het BAHD te plaatsen aan de dove zijde.

Deze proefschriftstudie is geconcentreerd tot de paediatrische patiënten populatie met dergelijke conductieve en gemengde gehoorverliezen. Het evalueren van de verkregen uitkomsten van deze BAHD behandeling bij die slechthorende kinderen is het hoofdonderwerp van deze proefschriftstudie. Een aantal nieuwe indicatie gebieden voor deze BAHD behandeling bij kinderen worden gevalueerd met daartoe geëigende en gangbare vragenlijsten als meetinstrument (kwaliteit van leven studies) en om zo de waarde van die behandelingen in maat en getal te kunnen uitdrukken.

Het plaatsen van de percutane Hoorschroef blijkt bij kinderen onder 5 jaar vaker gepaard te gaan met een hoger percentage van verlies van de Hoorschroef in vergelijking tot de groep oudere kinderen en de volwassenen. Met de invoering van de Softband in 2001 kwam als alternatief een transcutane toepassing voor handen om dit beengeleider hoortoestel aan het hoofd te helpen fixeren. Voor de

allerjongste groep kinderen werd dit al snel zo de voorkeursbehandeling om zo een operatieve plaatsing van de Hoorschroef in ieder geval enige jaren te kunnen gaan uitstellen. Nu is het aldus zelfs mogelijk geworden om deze groep kinderen al snel na de geboorte te kunnen gaan revalideren vanwege een aanwezig groot dubbelzijdig conductief gehoorverlies. Tegelijk kunnen zo de peri- en post-operatieve morbiditeit als gevolg van een percutaan te plaatsen Hoorschroef ontlopen worden.

Het is algemeen gekend dat een dubbelzijdig (conductief of gemengd) gehoorverlies op kinderleeftijd zonder een goede revalidatie leidt tot een aanzienlijke ontwikkelingsachterstand, onder meer in de taal- en spraakontwikkeling. Zoiets leidt weer tot gedragsstoornissen en uiteindelijk ook tot een lager scholingsniveau. Door onze recente gunstige ervaringen bij kinderen met een dubbelzijdige in plaats van alleen een enkelzijdige gehoorrevalidatie met BAHD's zijn wij zelf nog beter gaan begrijpen hoe essentieel een dubbelzijdige gehoorrevalidatie en daarmee een tweeorigheid voor het zich ontwikkelende kind is.

Volwassenen met een enkelzijdige volledige binnenoordoorfheid (SSD) blijken geholpen te kunnen worden met het plaatsen van een BAHD aan de zijde van het dove oor. Audiometrische metingen en subjectieve ervaringen gekwantificeerd met gestandariseerde vragenlijsten ondersteunen die conclusie. De effecten die op het gebied van de kwaliteit van leven bereikt worden zijn weliswaar wel geringer in vergelijking met de eerder al bereikte resultaten vanwege grote conductieve en gemengde gehoorverliezen. Tot nu is er echter nauwelijks enige ervaring met deze behandeloptie vanwege een eenzijdige binnenoordoorfheid bij kinderen. In dit proefschrift wordt beschreven dat de verzorgers van deze kinderen met een SSD en een BAHD behandeling een verbetering in de algemene gezondheid en in de kwaliteit van leven waar namen.

De gevolgen van een eenzijdig geleidingsverlies kan bij kinderen nogal variëren. Sommige kinderen presteren desondanks op school goed. Andere kinderen komen er door in de problemen. In deze proefschriftstudie - wat trouwens ook gebruikelijk is in de op dit punt voorhanden zijnde literatuur - richten wij ons op juist die kinderen met een eenzijdige geleidingslethorendheid die al in de problemen zijn geraakt. De ouders gaan op zoek naar hulp en vragen naar de mogelijkheden voor een revalidatie van dat gehoorverlies. Deze kinderen hebben dan al langere tijd een deprivatie van dat gehoor aan een zijde. Zij zijn in feite al

lange tijd functioneel eenorig. Een behandeling met een BAHD aan de aangedane zijde blijkt dan op korte termijn te leiden tot verbeterde kwaliteit van leven uitkomsten. Toch blijft de vraag wat dan toch de uitkomsten op de langere termijn zullen zijn en of toch niet al op een veel jongere leeftijd en zo meer succesvol zo een behandeling ingezet zou moeten gaan worden.

De literatuur meldt in toenemende mate het grote belang om tweeorig te mogen zijn en dit in vergelijking met de betekenis van eenorigheid. Het uiteindelijk te bereiken ontwikkelingsniveau, gemeten aan de resultaten van het uiteindelijk bereikte scholingsniveau, blijkt voor tweeorige kinderen hoger wederom in vergelijking met eenorige kinderen. Kinderen met een dubbelzijdig conductief gehoorverlies worden doorgaans vroegtijdig opgespoord, maar worden tot nu toe desondanks alleen enkelzijdig - veelal met een Softband en een BAHD - gerevalideerd. Deze proefschriftstudie toont, dat de toevoeging van een tweede BAHD contralateraal aan die Softband leidt tot een duurzame verbetering van de kwaliteit van leven uitkomsten. Deze resultaten benadrukken het belang om al vroegtijdig een dubbelzijdige gehoorrevalidatie te willen realiseren.

Het Birmingham University Medical Centre kent een eigen Kinderziekenhuis. Het paediatrische BAHD programma aldaar bestaat nu al weer zo een 20 jaar. Het geldt als een zeer gespecialiseerd tertiair centrum en een deel van de aldaar in behandeling zijnde kinderen hebben uitdrukkelijk behoefte aan een complexe multidisciplinaire zorg. Opmerkelijke craniofaciale anomaliën zijn bij deze kinderen niet ongevoel. De meeste chirurgische behandelingen bij kinderen kunnen eigenlijk alleen onder een algemene narcose worden uit gevoerd. Chirurgie om de percutane Hoorschroef bij kinderen te kunnen plaatsen geschiedt doorgaans in twee afzonderlijke operaties. Bij volwassenen gebeurt dit doorgaans in een operatieve sessie. Bovendien is het aantal revisie operaties vanwege de Hoorschroef bij kinderen vergeleken met volwassenen beduidend hoger. Dat betekent dus dat voor eenzelfde BAHD behandeling bij een kind meerdere algemene narcoses nodig zullen blijken te zijn. Met het al zo lang bestaan van een BAHD-centrum voor de BAHD-behandeling van kinderen in Birmingham is deze behandeling in Birmingham een tamelijk reguliere behandeling geworden en zo ook in aantal over de tijd fors toe genomen. Ten behoeve van de narcose wordt bij voorkeur gebruik gemaakt van een laryngeaal masker om zo een laryngeale en tracheale intubatie te helpen voorkomen. Dit alles ondanks de niet zo zeldzame problematiek van craniofaciale anomalieën bij deze patiëntgroep en daar mee een verhoogde moeilijkheidsgraad om aan deze kinderen een narcose te geven. Met

een laryngeaal masker blijkt een lage incidentie van complicaties voor te komen, maar hoe dan ook moet onder alle omstandigheden een open en veilige doorgankelijkheid van de luchtweg gegarandeerd kunnen blijven worden. Dat vereist de directe betrokkenheid van een hierin zeer gespecialiseerd Multi-disciplinair team. Dat argument alleen al is sterk genoeg, los van de andere benodigde expertises, dat de BAHD behandeling bij kinderen voorbehouden moet blijven aan dergelijke grote hoog gespecialiseerde medische centra.

Curriculum Vitae

Rupan Banga was born on 26th December 1974 in Nangal, India and grew up both in the Middle East and the UK. Her pre-university education was at St Christopher's School in Bahrain (1984-1991) and at Wakefield Girls High School in the UK (1991-1993). She studied Medicine at the University of Sheffield and graduated in 1998 having been awarded MBChB.

Her pre-registration house officer year was spent working at The Royal Hallamshire Hospital in Sheffield, and following this, she spent a further year working and travelling in Queensland, Australia.

On her return, she embarked upon Basic surgical training in Essex, Oxford and Nottingham (2001-2005) and during this time obtained the MRCS from the Royal College of Surgeons in Edinburgh.

Rupan was appointed onto the six year West Midlands Otolaryngology Specialist Registrar Training Program in 2005. During this time she has published and presented several papers on bone anchored hearing devices.

She was awarded the FRCS (ORL-HNS) from the Royal College of Surgeons in Edinburgh in 2009. She completed the training program and was awarded her Certificate of Completion of Training in 2011.

After completing her training, Rupan was awarded the Thomas Wickham Jones Otology Fellowship in Perth for a year with Professor Gunesh Rajan. Since her return to the UK, Rupan has been the Skull Base Fellow at Queen Elizabeth University Hospital Birmingham with Mr Richard Irving for six months and now has commenced as Consultant in the same department.

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Over the course of my education and career, there have been a number of friends, family and colleagues that have inspired, encouraged and motivated me to progress and achieve my goals. Similarly, this PhD Thesis would not have been possible without the support of so many individuals but there are a few who I would like to thank in particular for their guidance.

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-



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