BAHA

EVALUATION OF EXTENDED INDICATIONS SUCH AS MENTAL RETARDATION AND UNILATERAL HEARING IMPAIRMENT

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Ars longa, vita brevis. De kunst is lang, het leven kort. (Hippocrates)

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General introduction

General introduction

The bone anchored hearing aid (BAHA) was developed in Goteborg thirty years ago [1] This new type of bone conduction hearing aid has been commercially available in Goteborg since 1987 The Radboud University Nijmegen Medical Centre (UMCN St Radboud) started to apply the BAHA in 1988 Since then, over 1000 BAHAs have been implanted at the UMCN During almost thirty years of experience, the BAHA system has become a well-established treatment The BAHA proved to be a valuable option for hearing impaired patients with conductive or mixed hearing loss Over the years, multiple centres in different countries have started to apply this treatment technique BAHA treatment forms part of everyday practice in 30 out of 100 hospitals in the Netherlands Indications for application have gradually become broader

Previous Nijmegen BAHA PhD theses

The Nijmegen BAHA team has performed a great deal of research into the impact of the BAHA on patients with many different indications Audiologists and clinicians have worked closely together The promising results from our Swedish colleagues are supported by our own ascertainments Increasing experience with the BAHA and encouraging long-term results have led to new indications and their gradual implementation at the Nijmegen centre These results have contributed to FDA approval (USA) for application of the BAHA and to the worldwide popularity of the system In Nijmegen, continuous evaluation of various aspects of the BAHA has resulted in 4 PhD theses up till now The present BAHA PhD thesis is the fourth in Nijmegen In 2002, another PhD thesis based on the outcomes of the Birmingham BAHA Programme was defended in Nijmegen by Sunil Dutt In 2008, Mrs Ann Louise McDermott, also from Birmingham, will defend her BAHA PhD thesis on "The benefit and success of the Bone Anchored Hearing Aid" in Nijmegen All 6 theses present the results of BAHA fitting in the fields of clinical, audiological and patient outcomes

- 1994 E A M Mylanus, The Bone Anchored Hearing Aid, clinical and audiological aspects
- 1998 C T M van der Pouw, Bone anchored hearing, short and long-term results
- 2002 S N Dutt, The Birmingham Bone Anchored Hearing Aid Programme, Some Audiological and Quality of Life outcomes
- 2005 M K S Hol, BAHA, New indications and long-term patient satisfaction

- 2008 A McDermott, The benefit and success of the Bone Anchored Hearing Aid
- 2008 S Kunst, BAHA, Evaluation of extended indications such as mental retardation and unilateral hearing impairment

This fourth BAHA PhD thesis at the Radboud University Nijmegen Medical Centre is based on audiological and patient outcome aspects Evaluation of clinical results and patient outcome is of importance to obtain information about BAHA application in different groups of hearing impaired patients These further insights can be used to improve clinical practice and for the purpose of patient counselling

Physiology of bone conduction

The principle of bone conduction has been a study topic for many centuries In the 17th century, the first description was given of the phenomenon of bone conduction to aid patients with hearing impairment

Since the 19th century, bone conduction tests have been able to differentiate between conductive hearing loss and sensorineural hearing loss Different theories on the concept of bone conduction were put forward in the 20th century Many investigators have tried to understand this mechanism [2-6] Von Bekesy and Tonndorf played important roles in understanding and describing the physiology Von Békésy was the first to show that air conduction of sound signals and bone conduction pathways caused identical stimulation of the cochlea [2] This was brought up again recently in a paper by Stenfelt et al (2007) after they performed an extended version of the experiment described by Von Bekésy in 1932.[7]

The concepts of compression and inertia are the most commonly accepted explanations of how the cochlea is stimulated during bone conduction In 1966 Tonndorf stated that the mechanism of sound perception by conduction through bone cannot be explained by a single concept He described several factors and subfactors that all contribute in some way to stimulation of the cochlea Besides the inertia and compression concepts, Tonndorf hypothesized that vibrations in the bony and cartilaginous walls of the auditory canal were transmitted to the tympanic membrane which also contributed to stimulation of the cochlea [8] Although a large number of studies have been published on this issue many investigators are still trying to unravel the exact physiology of this phenomenon In Halifax, Bance et al investigated skull vibrations and the efficacy of bone conduction transducers [9] As mentioned above, Stenfelt et al published a recent paper on an extended version of the experiment described by Von Békésy [7] More

insight into the exact mechanism of bone conduction might lead to better implementation of the BAHA.

Osseointegration

The technique of titanium fixture implantation in bone emanated from dentistry.[10] In dental and craniofacial reconstruction, the percutaneous implant consists of a titanium fixture that becomes anchored in the skull due to osseointegration; afterwards, the percutaneous abutment can be attached to it. Certain circumstances are required to achieve the best osseointegration that guarantees long-term stability and the capacity to withstand load and stress.[11-14] Tjellström et al. were the first to combine the principle of osseointegration with the concept of direct bone conduction and hearing aids.[12] Successful osseointegration depends on several factors: material, design and surface of the implant. Nowadays a self-tapping titanium screw is commonly used. In addition, the surgical implantation technique is of importance. The success rate of standard BAHA implants is very high. Tjellström et al. (1995) reported 6.0 % implant loss in adults during a follow-up of 1-8 years.[15] Reyes et al. reported implant loss in 9 patients because of a lack of osseointegration and in 4 patients as result of trauma, thus a total of 13 cases out of 149 patients (11.5%) during 0-8 years of follow-up.[16] In 2006 Gillet et al. mentioned 2% implant loss during a follow-up of 0-8 years in a series of 63 patients.[17]

The fixture failure rate in adults was lower than in children.[15] In 100 children Granström et al. (2001) reported implant loss in 5.6%.[18] Davids (2007) reported a fixture failure rate of 10% in a group of twenty children younger than five years.[19] Lloyd et al. (2007) reported 26% implant loss in children from the Great Oromond Street BAHA programme in London. [20] McDermott et al. (2007) mentioned a 30% implant loss in children. [21]

Surgical procedure

Initially, a two-stage surgical procedure was applied to attain primary osseo-integration of the fixture, before any stress was applied with the percutaneous abutment. The first stage involved the installation of the titanium implant, which was left to osseointegrate for at least 3 months. In the second stage, the skin-penetrating abutment was placed. Nowadays a one-stage procedure is commonly used in adult patients.[15,22,23]

In young children, implantation of a titanium fixture is not straightforward. One reason is that the skull is still fairly thin (about 2 mm at the age of 2-3 years).[24] To achieve adequate fixation, the skull should preferably be at least 2.5 mm thick.

Another reason is that the structure of the bone of infants is not optimal for good osseointegration [25] Therefore, it is advisable to wait until the child is at least 2-3 years of age It has been suggested that a two-stage procedure leads to the best osseointegration This two-stage procedure is recommended in children of up to the age of about 10 years Nowadays it is no longer necessary to perform implantation before the age of 3 to 4 years owing to the introduction of the BAHA-softband Thus surgery can be postponed until the requirements concerning the thickness of the cortical bone and its composition has been met As the rate of implant loss is high especially in very young children, it may be advantageous to place a sleeping fixture during the first stage procedure to enable rapid continuation of BAHA use in the event of traumatic loss or implant failure [26]

To facilitate implantation a few changes have been made to the BAHA application procedure One of these innovations is the self-tapping implant which was based on experience with intra-oral implants [27] Recent research has shown that there was no difference in the level of osseointegration between the standard and self-tapping implants and that the implantation success rates were equal [27] Since 2004, a self-tapping fixture can be placed in a one-stage procedure with the abutment

Conventional bone conduction devices

In patients with conductive hearing impairment, reconstructive microsurgery of the middle ear is generally the preferred treatment A young age under the 6 years is considered to be a contraindication for elective surgery in children with major congenital ear anomalies In minor congenital ear anomalies surgery is contra-indicated in the age under 8-10 years So initially, treatment with a hearing aid is the only option Children and adults with inoperable major or minor congenital ear anomalies can be fitted with an air conduction hearing aid (ACHA), or a bone conduction hearing aid (BCHA) However, bilateral congenital anomalies of the pinna and aural atresia often make it impossible to fit an air conduction hearing aid In addition, recurrent or persistent otitis media or externa are contra-indications for an air conduction hearing aid because the mould blocks the ventilation of the external auditory canal In such cases, a bone conduction hearing aid is a good option

Typically, a conventional bone conduction hearing aid (BCHA) comprises a behind-the-ear device connected to a bone conduction transducer that is held in place by means of a steel band over the head, or mounted in a spectacle frame This transducer is positioned against the mastoid of the temporal bone with (considerable) static pressure to ensure correct functioning Variation in pressure

between the transducer and mastoid process is detrimental to speech recognition The constant pressure of the transducer can cause various complaints, such as local pain, skin irritation, headaches and furrowing of the squama in young children Another disadvantage is that the microphone and the vibrational transducer are positioned contralaterally, or at the level of the chest, which constitutes an unnatural listening condition Bone conduction via a spectacle frame does not have the latter disadvantage, but it does require the patient to wear glasses with a solid frame to generate sufficient static pressure This set-up has proved to be unsuitable for children Another drawback with the conventional bone conductor is that the patients suffer from social problems owing to unattractive appearance of the device [28]

The bone anchored hearing aid (BAHA)

The BAHA was designed in Goteborg as a special bone conduction device that aimed to deal with many of the above-described disadvantages It comprises a percutaneous titanium fixture anchored into the skull, a titanium abutment that is connected to the fixture and also penetrates the skin, plus a transducer that is connected to the protruding part of the abutment The vibrating transducer is coupled directly to the skull without any interference from intermediate tissue This percutaneous coupling is much more efficient than the transcutaneous coupling of the conventional bone conductors in the order of 10 to 15 dB [25] BAHA results proved to be superior to those obtained with conventional BCHAs When a BCHA is indicated, the BAHA is considered to be the first choice Similarly, in patients with severe bilateral conductive hearing impairment and an air-bone gap that exceeds 30 dB, the BAHA might be a better choice than conventional air conduction hearing aids (ACHAs) Mylanus et al concluded that if the air-bone gap was wider than approximately 30 dB, better results could be expected with the BAHA than with an ACHA [29]

BAHA sound processors

In 1987, the BAHA HC 100 became commercially available as the first experimental BAHA Since then, different types have emerged as a result of updating the HC 200 and the HC 300, better known as the BAHA Classic, in 1992 The BAHA Classic is suitable for patients with conductive or mixed hearing loss with a sensorineural hearing loss component of up to 30 to 35 dB HL

The BAHA Cordelle, a body-worn device, became available in 1998 This more powerful device can be applied to patients with a sensorineural hearing loss

component exceeding 35 dB HL up to approximately 60 dB HL [30] The hearing aid is worn partially at chest level

In the same year the BAHA Compact was developed It is smaller than the BAHA Classic and therefore has a more aesthetic appearance Maximum output is lower so it can only be used in patients with a sensorineural hearing loss component of 30 dB HL or less The BAHA Compact has proved to be less sensitive to interference from mobile phones

The successor of the BAHA Compact is the BAHA Divino (2005) This new type features a digital sound processor combined with a built-in directional microphone Protection against mobile phone interference has also been improved The Divino is suitable for patients with bone conduction thresholds of the indicated ear better than or equal to 45 dB HL Additionally, patients with unilateral sensorineural hearing loss of the indicated ear and normal contralateral hearing (BAHA-CROS) will also benefit from the Divino sound processor Sound quality proved to be significantly better with the BAHA Divino [31]

In 2007, the BAHA Intenso was introduced as the most recent type It has extra power and an advanced digital signal, which makes it easier to use and more comfortable in background noise This new processor bridges the gap between the Divino and Cordelle Preliminary test results with this new BAHA are very promising

BAHA Softband

Transcutaneous application of the BAHA was initiated and introduced in 2002 in Nijmegen to provide young children with early access to auditory stimuli The so-called BAHA Softband comprises an elastic band around the skull that holds the BAHA in place and enables transcutaneous transmission of sound via bone conduction This temporary solution for children awaiting implantation is more practical and comfortable to wear than a conventional steel headband In a study by Hol et al , the BAHA softband proved to be a valid intervention in two children with bilateral congenital aural atresia Their speech and language development progressed in accordance with their cognitive development [32] Verhagen et al (2008) evaluated more children with a longer follow-up and again showed the additional value of the BAHA softband [33]

First indications for BAHA

Initially, the target group consisted of patients with mixed or conductive hearing loss who could not be fitted with a behind-the-ear or in-the-ear hearing aid and in whom a conventional bone conduction hearing aid had proved to be inadequate

and impossible to fit. Furthermore, microsurgery was not expected to offer a reasonable alternative, or achieve a dry ear, to enable treatment with an air conduction hearing aid. The first series of exclusion criteria was described in the thesis by Mylanus (1994):

1. Age of younger than 10 years
2. Pure sensorineural hearing loss
3. Conventional hearing aids provide satisfactory results
4. Mixed hearing loss of greater than 65 dB HL at the frequencies 0.5, 1 and 2 kHz
5. Lack of personal motivation or acceptation problems

Over the past thirty years, the exclusion criteria have gradually been adjusted and redefined.

Extension of the indications and new patients groups

BAHA in children

Owing to the requirements concerning adequate thickness of the cortical bone and its composition implantation was not done in young children before the age of 2 to 3 years. Nowadays, as a result of the availability of the BAHA-softband, implantation can usually be postponed until the age of 4-5 years. After surgery, the percutaneous titanium implant improves the supported hearing level by about 10 dB compared to a transcutaneous set-up.

The BAHA Softband was developed for very young patients with bilateral conductive hearing loss, who were too young for implantation. A study by Yoshinaga et al. confirmed the importance of early acces to auditory stimuli, preferably before the age of 6 months. Delayed intervention might lead to permanent language deficits.[34] They reported that language test results in the children who had received hearing aids before the age of 6 months were significantly better at the age of 3 to 4 years than those of children who had been fitted later in life. A conventional bone conduction device on a steel headband is not popular so early in life and it can only be applied for limited periods. Therefore, the BAHA Softband is a valuable alternative in terms of comfort and utility before the age of 6 months. Further studies have shown that it is very effective to apply bilateral BAHAs with this Softband. At a later date, bilateral percutaneous implants will result in even better sound transmission due to the percutaneous coupling instead of the transcutaneous set-up with the BAHA Softband.

Unilateral congenital conductive hearing impairment

In the past, unilateral hearing impairment was not considered to be a very significant handicap, because speech and language were presumed to develop appropriately. Furthermore, no suitable surgery or hearing aid was generally available, which might have contributed to this view. However, in 2004, Lieu et al reviewed a number of studies[35-38] and found that speech and language development were often delayed in children with unilateral hearing impairment and that it was unclear whether they could really "catch up" as they grew older.[39] School age children with unilateral hearing impairment appeared to have increased rates of school year failure, they needed additional educational assistance and tended to display behavioural problems in the classroom. It is therefore very likely that such children would benefit from early hearing aid fitting.[39] The BAHA is a valuable option that is worth consideration in this respect.

The BAHA in patients with moderate mental retardation and conductive hearing loss

The BAHA system was developed as an alternative for patients with conductive hearing loss who could not be fitted with ACHAs.

Research has shown that the BAHA is the best bone-conduction hearing aid available and it is considered to be particularly suitable for the treatment of conductive or mixed hearing impairment.[25 40-42]

It is still not clear at which age it is best to switch a child from transcutaneous to percutaneous application in order to optimise their personal intellectual development.[33] Based on only a few reports, the age of 4 years is currently considered to be justified.

After implantation, the percutaneous titanium fixture requires regular personal care, including close attention to the surrounding skin. Patients with moderate mental retardation were initially excluded from BAHA treatment, because of doubts about compliance with caring for the percutaneous implant and the surrounding skin.[25] So there was a suspicion to encounter an elevated fixture extrusion rate in these patients. Social isolation is a common problem in patients with mental retardation combined with hearing loss. To enable an adequate level of social communication, optimal hearing aid fitting is necessary. The BAHA forms a good solution in patients with conductive hearing loss and recurrent otitis and may also be beneficial in patients who have these problems in combination with moderate mental retardation.

In view of the encouraging long-term results of BAHA application, the first persons with moderate mental retardation entered the BAHA treatment programme in

Nijmegen in 1996 Their results were published in 2006 and 2007 [43,44] Over a period of 8 years, 22 patients with moderate mental retardation have received a BAHA The majority of patients have Down's syndrome In this thesis, we evaluated whether the BAHA is a good option in hearing impaired patients with moderate mental retardation [45] We also explored whether the indications for BAHA application might be extended further

In 2006, Sheehan et al studied BAHA application in 43 patients with Down s syndrome [45] Their multicentre retrospective study showed a high level of satisfaction with the BAHA among the patients, parents and care providers No subjective evaluations were made with health-related questionnaires

The BAHA in patients with unilateral acquired and unilateral congenital conductive hearing loss

It is still a challenge to achieve binaural hearing at an early age in patients with unilateral congenital conductive hearing impairment Conventional microsurgery for minor congenital middle ear anomalies is successful in about 80% of the cases, but surgery is usually postponed until the age of 10 years [46,47] When major congenital ear anomalies are present, including atresia of the bony canal, microsurgery is only an option in cases with minimal involvement of the ear canal, such as class I or IIa atresia (classification Altmann-Cremers) [47] However, very few of the patients with unilateral congenital ear canal atresia have these less severe classes and form candidates for microsurgery Furthermore, this type of surgical intervention is not usually performed until the age of 6 years in children with ear canal atresia, or until the age of 10 years in children with isolated congenital ossicular chain anomalies Therefore, other options have to be found, e g early BAHA fitting with the softband

In 1994, Wilmington et al [48] measured binaural hearing skills in patients with unilateral congenital conductive hearing loss before and after successful reconstructive surgery On a group level, there were significant improvements in localization abilities of their patients However, the post-surgery scores were still poorer than those found in subjects with normal hearing The same tendencies were also visible in the directional hearing experiments but remarkably, about one third of the patients had a fairly good score before surgery Apparently, these patients had learnt how to compensate for this deficit In 1994, Snik et al also studied the outcome of successful surgery for congenital unilateral conductive hearing impairment They reported that the results were within the normal range 1 year after surgery [46]

An alternative to surgery is to fit a conventional ACHA, especially when the hearing impairment is less than 40 dB However, in patients with complete atresia of the bony ear canal, this is not an option Furthermore, when the unilateral conductive hearing impairment is about 60 dB, very high levels of amplification are needed, which cannot always be provided by an air conduction hearing aid, owing to feedback problems A better solution therefore might be a bone conduction hearing aid, such as the BAHA, as it avoids most of the drawbacks of the conventional transcutaneous bone conduction devices [40]

The BAHA restored binaural hearing successfully in patients with unilateral acquired conductive hearing loss [49 52] In 2005, Hol et al reported significantly improved sound localization, speech recognition and subjective benefit in 18 patients with an acquired unilateral air-bone gap [51] Chasin and Wade (1998) observed improvement in binaural summation and in masking level differences in 6 patients with unilateral conductive hearing impairment due to unilateral congenital atresia [52] In a group with mixed and conductive unilateral hearing impairment (congenital and acquired), Wazen et al (2001) reported that speech recognition performance in the BAHA-aided condition was comparable with the patient's best score in the unaided condition [49] Directional hearing function was not measured in this study

In an earlier pilot study, Snik et al (2002) found that although the BAHA effectively improved directional hearing in the patients with acquired unilateral conductive hearing loss, the results were ambiguous in two patients with unilateral congenital conductive hearing impairment [50] Directional hearing tests after BAHA fitting did not show any improvement in these two patients, because their localization abilities were already good in the unaided condition There was no clear explanation for this phenomenon

Intervention is advisable in children with unilateral hearing loss, because they have a significantly higher risk of developmental delays than their peers with normal hearing [39] However, studies on conventional hearing aid use in such children showed poor compliance (e g Davis et al , 2002) [53] In their position statement on unilateral hearing loss, the Paediatric Workgroup (Bess et al , 2000) stated that amplification should be considered case-by-case, based on the child's development and communication needs [54]

Quality of life aspects after medical interventions are receiving more and more attention In a study by Arunachalam et al (2001) on changes in quality of life following BAHA fitting, the Glasgow Benefit Inventory demonstrated clear improvements in the subgroup of patients with bilateral congenital atresia

(n=12) [58] In 2004, Mc Larnon et al found that the patients with congenital ear disorders (n=10) were likely to experience the most benefit from a BAHA [59]
Besides that, in patients with bilateral conductive hearing loss, BAHA application proved to be an effective means to restore binaural hearing [55 57]
These promising audiometric and subjective results of the BAHA in patients with bilateral conductive hearing loss encouraged us to extend the indications for BAHA application
In 1998, we started to use the BAHA system to treat patients with severe acquired or congenital unilateral conductive hearing impairment who did not have any other treatment options Audiometric and subjective assessments on a prospective group of 18 consecutive patients with unilateral acquired conductive hearing loss showed restoration of binaural hearing and high patient satisfaction [50 51]
Subjective outcomes of BAHA fitting in 20 patients with unilateral congenital conductive hearing impairment are included in this PhD thesis (chapter 3 1) together with recent reports in the literature [60 62]

The BAHA in patients with single sided deafness
In patients with unilateral sensorineural deafness, a BAHA positioned behind the deaf ear works as a transcranial CROS (Contralateral Routing of Sound) device Sounds received by the BAHA system, are transmitted by bone conduction to the functional contralateral cochlea Application of the BAHA CROS in unilateral sensorineural deafness has been studied at several centres [63-69] Overall, these studies reflected that the BAHA CROS effictively lifted the head shadow, which was advantageous in specific listening situations, for example at the dinner table, while driving a car, or at a meeting when the speaker was on the impaired side A conventional CROS or a BAHA CROS cannot achieve stereophonic hearing This was corroborated by sound localization results that did not differ from chance level [64 66 68 69]
Despite the lack of documented changes in sound localization abilities, questionnaires that measured patients' opinions showed substantial subjective benefit and indicated considerable satisfaction [63-66 70] Subjective reports of improved sound localization abilities after BAHA implantation may be due to aleviating of the head shadow effect Any lack of improvement in sound localization abilities was counterbalanced by the positive impact of better communication in several specific situations

Before implantation, counselling of the patients is of great importance Lifting the acoustic head shadow alone might be a major advantage to some patients with

certain life-styles and occupations. Prior to implantation, a trial should be arranged in which a BAHA is positioned against the mastoid of the deaf ear using a steel headband. At the Nijmegen clinic, about 25% of the patients who applied for a BAHA CROS did not experience sufficient benefit and stopped using the steel headband set-up during the trial period. Despite previous counselling, their expectations, especially concerning directional hearing, were probably too high. A recent study by Andersen et al. showed that half of the patients with unilateral hearing loss after acoustic neuroma surgery accepted a trial period with the BAHA on a steel headband. Ultimately, 25% were sufficiently satisfied to apply for implantation surgery and BAHA fitting.[71]
This thesis describes the outcome of BAHA CROS application in 56 patients, which is the largest series in the literature. A review of the most recent literature on BAHA CROS application is given in chapter 4.1.[62]

Scope of this thesis

The scope of this thesis covers 3 different themes: moderate mental retardation, unilateral congenital conductive hearing loss and single sided deafness.
In *chapter 2*, the combination of mental retardation and conductive hearing loss was studied as a new indication. Initially, these patients were excluded from BAHA treatment, because it was doubtful whether they or their care providers, would be able to cope with the necessary daily care of the percutaneous titanium implant. In 1996 after almost 10 years of experience with BAHA application at the Radboud University Nijmegen Medical Centre, the first patients in this special group entered the BAHA programme. Selection was made of 22 patients with moderate mental retardation and conductive or mixed hearing loss. Slightly more than a half of these patients had Down's syndrome. The aim of this study was to asses the implantation results, audiological results (chapter 2.1) and subjective benefit (chapter 2.2). The Patients' family or care providers involved in daily care at institutes, filled out the Glasgow Children's Benefit Inventory (GCBI) and the Listening Inventory For Education (LIFE).
The second theme (*chapter 3*) consists of an evaluation of patients with congenital unilateral conductive hearing loss. Application of the BAHA system to enable binaural hearing was succesful in patients with unilateral conductive hearing loss with acquired etiology.[49-52] Hol et al. reported significant improvements in sound localization, speech recognition and subjective benefit in 18 patients with an acquired unilateral air-bone gap.[51] In an earlier pilot study performed in 2002, Snik

et al found ambiguous results in two patients with congenital unilateral conductive hearing impairment, whereas the BAHA had effectively improved directional hearing in the patients with acquired unilateral conductive hearing loss [50] Directional hearing did not show any improvement in the two congenital patients after BAHA fitting, because their localization results in the unaided condition were unexpectedly high It remained unclear why the existing localization performance of these patients was so good

To find out whether this is a consistent finding and to gain more insight into audiological performance, we obtained test results from a larger group of congenital patients Over a period of 8 years, 20 patients with unilateral congenital conductive hearing impairment have been fitted with a BAHA Audiometric (chapter 3 1) and subjective (chapter 3 2) evaluations were done Speech discrimination in noise and directional hearing scores were recorded in a test setting Subjective evaluations were made using 2 different outcome measures the GCBI in the children and the Intern Outcome Inventory for Hearing Aids (IOI-HA) in the adults To gain more insight into sound localization abilities after BAHA fitting, the adults and children filled in appropiate parts of the Speech Spatial and Qualities of Hearing Scale (SSQ)

The third theme is described in *chapter 4,* regarding the use of the BAHA in patients with unilateral profound sensorineural hearing loss Traditionally the audiological approach consists of fitting a contralateral routing of sound (CROS) hearing aid As the interaural attenuation of sound conducted by bone is small with the BAHA, it can be used a transcranial contralateral routing of sound (CROS) device The first experience of the Nijmegen BAHA group with the BAHA CROS device was described in the thesis by Hol (2005) Contralateral routing of sound with the BAHA proved to be possible and the benefit of reducing the head shadow effect was confirmed with speech recognition measurements However, sound localization results were poor which meant that the BAHA CROS was unable to restore binaural hearing Subjective outcome measurements generally indicated considerable satisfaction

As it was not yet clear whether patients with unilateral sensorineural hearing loss benefited from BAHA application, we extended the previous group of 29 patients to 56 cases This enabled the evaluation of several different groups

In *chapter 4 2* the results are presented of a randomized controlled trial in which 10 adult patients with unilateral total inner ear deafness compared the conventional CROS, the completely in the canal hearing aid (CIC) and the BAHA CROS

In *chapter 5* a general discussion is given In thirty years of experience with the BAHA system, many BAHA studies have been performed Still some unanswered questions keep numerous researchers interested, such as the restoration of binaural hearing, directional hearing in patients with congenital hearing impairment, the plasticy of the brain after the restoration of binaural hearing, why some patients do not apply for implantation after a trial period with a BAHA on a steel headband, which patient characteristics predict a positive outcome after implantation, cost–benefit ratios and recent progress in the treatment of hearing impaired patients

Over the past few years, the technical options have broadened extensively The introduction of partially implantable hearing aid systems or middle ear implantations has opened the field to discuss choices between different amplification options Semi-implantable hearing aids as the Vibrant Soundbridge and otologics MET and the cochlear implant as a full-implantable hearing aid, are common used Such new developments have their own focus and will take a clear position in relation with the BAHA This also applies to the application of the Vibrant Soundbridge in the round window in patients with a large bilateral sensorineural component, mixed hearing loss or radical cavities, because it provides greater benefit than the present BAHA-Cordelle The results of cochlear implantation have been improved again especially because it is now within our scope to preserve the remaining inner ear function Those new techniques can be seen as additional audiological treatment options, or there maybe overlap with some BAHA indications On the other hand, the BAHA is still undergoing development, so new types with stronger effects and new design can be expected Evaluation of the BAHA should continue, in order to demonstrate the value of this treatment and to maintain its strong place among all the other options The BAHA in his present form has proved to be consistent and beneficial

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2.1

Rehabilitation of patients with conductive hearing loss and moderate mental retardation by means of a bone-anchored hearing aid

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Abstract

Objective: To evaluate whether the bone-anchored hearing aid (BAHA) can be applied successfully to patients with conductive hearing loss and moderate mental retardation.

Study Design: Retrospective clinical evaluation.

Setting: Tertiary referral centre.

Patients: Twenty-two patients with congenital moderate mental retardation and conductive or mixed hearing loss, selected to receive a BAHA at the University Medical Centre Nijmegen, the Netherlands. Four of them were fitted despite a limited air-bone gap.

Intervention: Rehabilitative BAHA application.

Main outcome measures: Implantation results, skin reactions and audiological data were evaluated over a mean follow-up of 36 months.

Results: All the patients were still using the BAHA 7 days a week and for more than 8 hours a day after a follow-up period between 5 and 96 months. Two implants (9%) were lost due to insufficient integration but were reimplanted successfully. With the BAHA, mean free-field thresholds showed a clear mean improvement of 9 dB compared to the previous hearing aid. Considerable improvements in daily activities were seen in at least 5 patients.

Conclusions: Moderate mental retardation should no longer be considered as a contra-indication for BAHA application. Although implant loss was low, extra attention may be required from the personal care providers to maintain the percutaneous implant. The BAHA was well-accepted by the patients with moderate mental retardation and was being used for most of the day. Implementation of the BAHA as hearing aid treatment in patients with moderate mental retardation proved to be sufficiently effective and may have strongly positive effects on activities at school or at work

Introduction

The BAHA system was developed in Göteborg as an alternative option for patients with conductive hearing loss who could not be fitted with air-conduction hearing aids (ACHAs). Since 1987 it has been introduced successfully in many other countries. Research has shown that the BAHA is the best bone-conduction hearing aid available and it is considered to be particularly suitable for the treatment of conductive or mixed hearing impairment.[1-4] This semi-implantable bone-conduction hearing aid is coupled to a percutaneous titanium screw implanted in the mastoid behind the auricle. Regular personal care is required for the percutaneous titanium fixture, including close attention to the skin around the implant. Awareness of the implant was considered to be essential in this care. When the BAHA system was first introduced, there was concern about the long-term outcome of the percutaneous titanium implant, because a too high extrusion rate was expected. Initially, it was doubted whether moderately mentally disabled patients and their care providers would be able to cope with the care needed to avoid loss of these implants.[3] Therefore, this group was excluded from BAHA treatment.[4]

Long-term outcomes of the percutaneous titanium implant were so good that after almost 10 years of experience with BAHA system application in Nijmegen in 1996, the first group of moderately mentally disabled patients were accepted for this treatment. Over a period of 8 years, 22 moderately mentally disabled patients have received this treatment at the Nijmegen University Medical Centre. The majority of patients had Down's syndrome. Difficulties were expected with cleaning the skin around the abutment and the use of the BAHA device in this patient group, so good follow-up was considered important. These patients were seen regularly at the outpatient clinic every 4 months to begin with and later at least yearly, with one of their care providers. During these check-ups, the stability of the implant and the surrounding skin were monitored. Skin reactions were classified according to the clinical system of Holgers et al.[5]

To the best of our knowledge, this is the first study on BAHA application to patients with moderate mental retardation. The aim of this study was to evaluate whether the BAHA is a good option for patients with moderate mentally retardation and to explore whether the indications for BAHA application can be extended.

Methods

Patient characteristics

A total of 22 successive patients with congenital moderate mental retardation and conductive or mixed hearing loss were selected for BAHA implantation at the University Medical Centre Nijmegen. The majority of these patients had Down's syndrome (12 patients), while the others had different forms of moderate mental retardation. Mean age at the time of implantation was 36 years (range 7-73 years). Mean age at the time of this study was 39 years (range 10-74 years). Time of implantation varied from July 1996 to June 2004. Follow-up varied from 5 months to 96 months (mean 36 months). The female: male ratio was 1.75: 1 (14 women, 8 men). An overview of the patient characteristics is presented in Table 1.

Table 1. Patient characteristics at the time of follow-up

Patient	Sex	Age (yrs)	Aetiology mental disability	Indication for BAHA	Living situation	Working situation
1	M	45	Down's syndrome	OME	Surrogate family home	DCC
2	F	22	Down's syndrome	COM, discomfort	Sheltered housing	Sheltered workshops**
3	M	41	Down's syndrome	COM	Family	DCC
4	F	24	Down's syndrome	COM, discomfort	Independent	Other*
5	F	31	Unknown	COE, discomfort	Independent	DCC
6	F	55	Down's syndrome	COE	Surrogate family home	Sheltered workshops
7	F	56	Unknown	COM	Independent	DCC
8	M	75	Unknown	COE	Home for the mentally handicapped	DCC
9	F	35	Unknown	COE	Surrogate family home	Supported employment
10	M	10	Grouchy syndrome	Atresia IIA	Parents	Special education
11	F	51	Down's syndrome	COE	Home for the mentally handicapped	DCC
12	F	66	Unknown	COM	Independent	None
13	M	64	Down's syndrome	COM	Home for the mentally handicapped	DCC
14	M	38	Down's syndrome	Discomfort	Surrogate family home	Supported employment
15	M	10	Down's syndrome	OME	Sheltered housing	Special education
16	M	22	Down's syndrome	COM	Parents	Other ***
17	M	30	Down's syndrome	COM	Parents	Supported employment
18	F	34	Unknown	COE, discomfort	Sheltered housing	DCC
19	F	21	Unknown & BOR syndrome	COE	Surrogate family home	Other ***
20	F	35	Kartegener syndrome	COM	Independent	Supported employment **
21	F	27	Down's syndrome	COE, discomfort	Surrogate family home	DCC
22	F	57	Unknown	COM	Surrogate family home	Sheltered workshops

Abbreviations M male, F female; OME otitis media with effusion, COM chronic otitis media; COE chronic otitis externa; DCC day care centre for the mentally handicapped

* Other. independent work, catering industry; independent work, gift shop; voluntary work

** Patients 1-2-16-19-20 (n=5) started work after receiving a BAHA, owing to vast improvement in social communication

All the patients had binaural hearing and received the BAHA unilaterally near the ear with the best bone-conduction thresholds. Except for one case, who had never worn a hearing aid, they had been using various types of conventional hearing aid (CHA) before implantation: a conventional air-conduction hearing aid (ACHA) n=14, a conventional bone-conduction hearing aid (BCHA) n=6 (3 with hearing spectacles) and 1 patient had been using an ACHA and a BCHA together. Table 2 presents hearing device data and the patient implant characteristics. Despite a limited air-bone gap (< 15 dB), 4 patients (nos 7, 8, 12, 18) with mixed hearing loss received a BAHA. The standard BAHA Classic was fitted in 11 patients, the more powerful BAHA Cordelle (a body-worn device) in 6 patients and a BAHA Compact, which is somewhat less powerful than the Classic, in 5 patients.

Table 2. Patient implant characteristics and hearing device data at the time of implantation Thresholds (on the side of implantation) are averaged at the frequencies 0.5, 1, 2 and 4 kHz; PTA (dB).

Patient	Age (yrs)	PTA BC	PTA AC	Previous HA	BAHA type	Implant side
1	44	41	64	ACHA	Classic	AD
2	20	23	76	BCHA	Compact	AS
3	33	45	76	ACHA	Cordelle	AD
4	16	18	35	BCHA	Compact	AD
5	29	40	80	BCHA*	Classic	AS
6	54	39	70	ACHA	Classic	AD
7	54	50	59	ACHAs	Cordelle	AS
8	73	59	73	ACHAs	Cordelle	AS
9	34	33	64	ACHAs	Classic	AS
10	7	6	46	None	Compact	AD
11	50	36	58	ACHAs	Classic	AS
12	65	46	60	ACHA & BCHA	Cordelle	AD
13	62	58	105	ACHA	Cordelle	AS
14	37	31	85	BCHA*	Classic	AS
15	9	15	48	ACHA	Compact	AD
16	18	44	71	ACHAs	Cordelle	AS
17	30	51	100	ACHAs	Classic	AS
18	34	36	51	BCHA*	Classic	AD
19	19	20	80	ACHAs	Compact	AD
20	27	21	45	ACHA	Classic	AD
21	21	25	49	BCHA	Classic	AD
22	51	36	74	ACHAs	Classic	AD

Abbreviations: BC bone-conduction, AC air-conduction, HA hearing aid, ACHA air-conduction hearing aid, BCHA bone-conduction hearing aid; BAHA bone-anchored hearing aid, AD auricula dextra; AS auricula sinistra

Note patient 10 had no previous HA; * Hearing spectacles

To gain insight into the number of hours of daily BAHA use and into problems with cleaning the percutaneous titanium implant in this special patient group, the

patients and their care providers were asked to answer the first 3 hearing aid related questions of the adapted Nijmegen questionnaire.[6] Two questions were added about their living and occupational situations.

Audiology

Audiological evaluation was performed using standard procedures and equipment, including play-audiometry. Pure-tone average air- and bone-conduction thresholds at 0.5, 1, 2 and 4 kHz were calculated from the pre-operative audiogram. About 6 weeks after BAHA fitting, aided free-field thresholds were collected and the maximum phoneme score (MPS), defined as the highest percentage of correctly repeated phonemes, was derived from the free-field speech recognition-intensity function (speech audiogram).[3] The phoneme score at 65 dB was also calculated from this speech audiogram. Similar measurements had been performed with the previous hearing aid prior to implantation, but not consistently. Results were available on 16 patients.

To analyze the results a two-tailed non-parametric test was used. $P< 0.05$ was chosen as level of significance. The results were computed using the SPSS software package (version 12.0; SPSS Inc, Chicago, Ill).

Results

All 22 patients were using their BAHA 7 days a week and for more than 8 hours a day. Over half of the care providers (59%, n= 13) reported that they did not have any problems with cleaning the abutment; 27% (n=6) faced difficulties sometimes and 14% (n=3) had actual cleaning problems.

In 20 patients, there had been no difficulties with osseointegration of the implanted screw. A total of 2 implants (9%) were lost owing to insufficient integration. One case developed infection of the skin around the implant, possibly as a result of psoriasis capitis at time of implantation, whereas the other case did not show any signs of infection. These 2 patients with lost implants we reimplanted successfully. Time between first implantation and loss was 4 months in both cases. Replacement took place after 1 month and 5 months, respectively. Since reimplantation, fixation of the percutaneous screw has remained stable in both patients during follow-up periods of 5 months and 87 months, respectively. In 1 other patient, only the abutment became loose and was lost at 18 months follow-up. A new abutment was immediately placed on the implant. During follow-up, adverse reactions were observed on the surrounding skin. No side-effects

occurred during follow-up in 14 patients Skin reactions around the implant were seen in 3 patients (2 of grade 1 and 1 of grade 3) Movement of the abutment in relation to the skin did not occur in any of the patients Relatively thick skin developed around the abutment in 5 patients One patient underwent surgical revision of the skin thickness and another patient received a larger abutment (8 5 mm instead of 5 5 mm) which formed a sufficient solution

Indications to receive a BAHA could be divided into 3 categories in these patients first, chronic otitis external combined with middle ear involvement (air-bone gap), second, sequelae of chronic otitis media that contraindicated the use of a conventional ACHA, third, the same etiological background as categories 1 and 2, but in addition severe pain, skin irritation or headaches due to the pressure of the BCHA against the skin of the temporal bone

The answers of the care providers to the questions about the living situation and working situation showed remarkable improvements in daily activities in at least 5 patients The work supervisors reported that these 5 patients had become able to work after receiving a BAHA, due to improvements in communication and thus social functioning

In the total group of patients, 2 were receiving special education, 7 had supported employment or sheltered workshops, 9 patients were visiting a day care centre for the mentally handicapped and 3 had independent work One patient was not involved in an occupational situation

Audiology

The pure tone average air- and bone-conduction thresholds at 0 5, 1, 2 and 4 kHz are presented in Table 2 One patient received a BAHA because of therapy resistant external otitis as a result of occlusion of the ear canal by the earmould of her ACHA, despite a small air-bone gap (9 dB) Mean air- and bone-conduction thresholds and mean free-field thresholds obtained with the BAHA are illustrated in Figure 1 The figure suggests that the BAHA system is effective in this patient group Mean free-field thresholds with the BAHA approached the mean bone-conduction thresholds Data on free-field thresholds with the previous hearing aid were only available in 9 patients Figure 2 presents the mean data of these 9 patients, including the free-field thresholds obtained with the previous device This figure shows better thresholds with the BAHA than with the previous device Mean free-field thresholds showed a mean improvement of 9 dB with the BAHA compared to the previous hearing aid However, this clear difference was not statistically significant (P= 0,098)

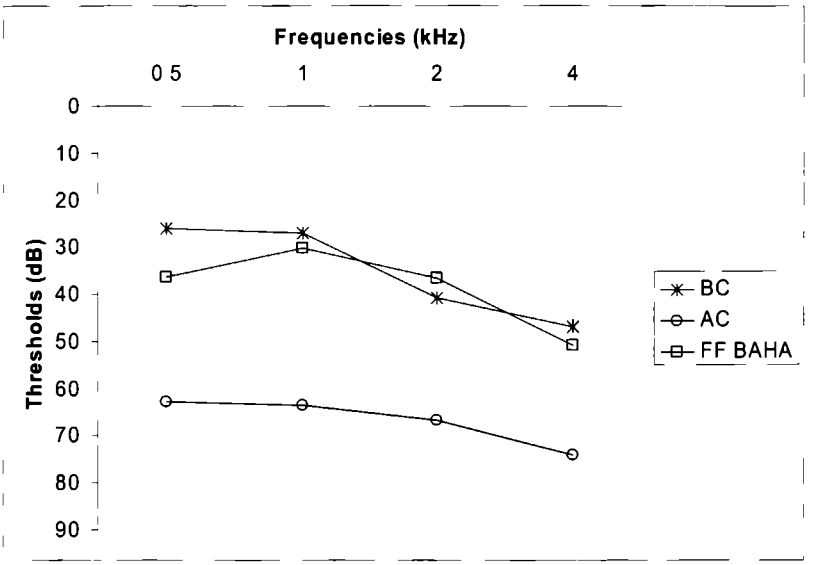

Figure 1. Mean air-conduction (AC) thresholds bone-conduction (BC) thresholds and mean free-field thresholds measured with the BAHA (FF BAHA) as a function of frequency (n=22).

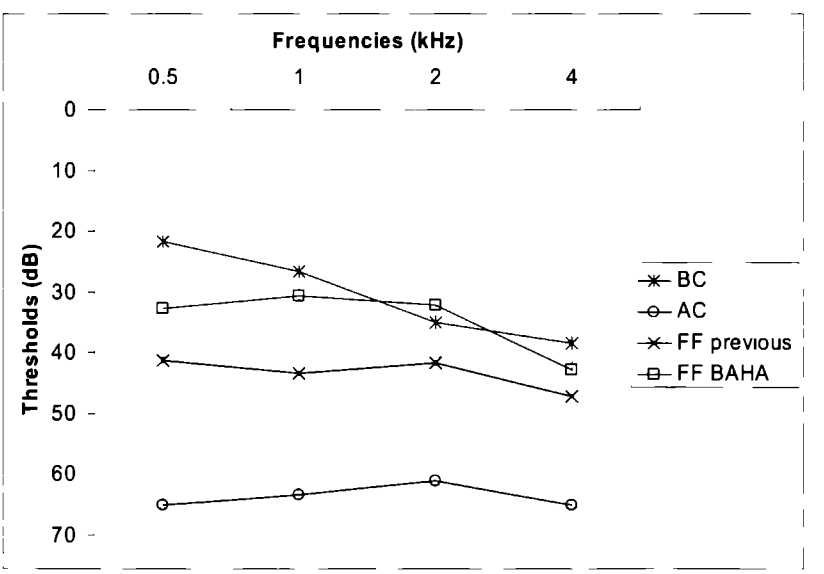

Figure 2. Mean air-conduction (AC) thresholds, bone-conduction (BC) thresholds and mean free-field thresholds measured with the previous device (FF previous) and with the BAHA (FF BAHA) as a function of frequency in the 9 patients with complete data sets.

Table 3 gives an overview of individual data on mean free field thresholds with the previous device and with the BAHA, speech recognition scores at 65 dB and maximum phoneme scores. In 5 patients, speech testing was difficult due to the severity of mental retardation.

Table 3. Audiometric data on the patients

Patient	Free field		65 dB		MPS	
	Previous	BAHA	Previous	BAHA	Previous	BAHA
1	n.a.	34	n a	85	n.a.	100
2	33	31	n a.	70	n.a.	70
3	53	43	35	85	n.a.	n.a.
4	31	26	95	100	100	100
5	n.a.	50	n.a.	n.a.	n.a.	n a.
6	n.a.	43	n a	n.a.	n.a.	n.a
7	n.a.	46	20	65	90	90
8	n.a.	n.a.	n a	65	n a	65
9	n.a.	38	n.a.	80	n.a.	80
10	60	25	n a.	90	n.a.	90
11	n a.	30	n.a.	n.a	n.a.	n.a
12	35	46	95	90	100	90
13	n.a.	50	n.a.	50	n.a.	70
14	60	43	0	70	80	95
15	n.a.	40	n.a	n a.	n.a	n.a
16	45	34	n.a	n a.	n.a.	n.a
17	n.a.	58	70	65	n.a	80
18	n.a	46	n.a	70	n.a.	100
19	n a.	33	100	90	100	100
20	n.a	29	100	90	100	100
21	30	35	85	90	n.a	n.a
22	41	29	55	80	95	n.a

Mean free-field thresholds averaged at the frequencies 0 5, 1, 2 and 4 kHz (dB) obtained with the previous hearing aid and with the BAHA Speech recognition scores (%) at 65 dB and at their maximum Abbreviations: MPS maximal phoneme score, n.a. not available

Individual speech recognition scores at 65 dB were compared to mean speech recognition scores from a control group of all the patients fitted with a BAHA at the Nijmegen University Medical Centre with available measurements (n=576). The data presented in Figure 3 show that the results of the patients with mental retardation were well within the range of the controls. Thus it appeared that the performance of the BAHA users with moderate mental retardation was comparable with that of the whole group of BAHA users.

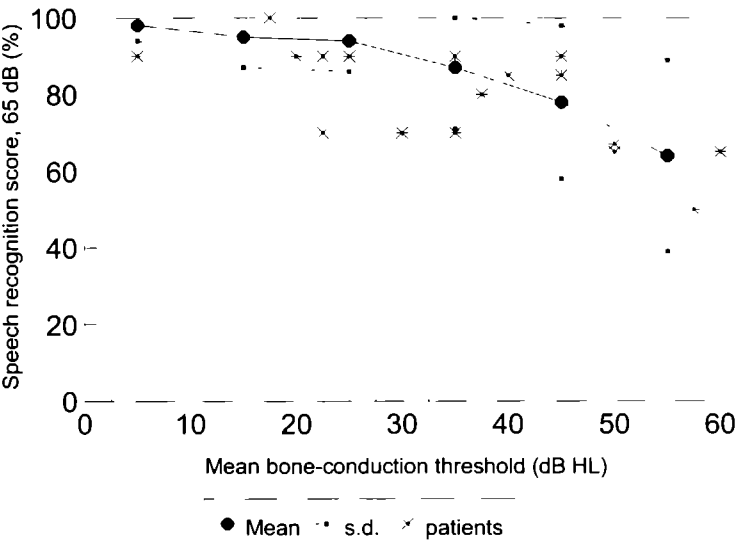

Figure 3. Speech recognition scores at 65 dB (%) with the BAHA (n=17) compared to mean data from a control group of BAHA users (n=576) versus the mean bone-conduction threshold averaged at the frequencies 0.5, 1, 2 and 4 kHz (dB). Control group data are presented as mean data ± 1 s.d. as a function of the mean bone-conduction thresholds divided classes of 10 dB wide.

Discussion

Nowadays the BAHA system is a well-established treatment in selected cases. Over the past decades the indications for the BAHA have been extended widely. The BAHA has also been approved by the FDA in the United States for adults and children. Mylanus et al.[7] reported that the BAHA proved to be an effective hearing aid for patients with conductive or mixed hearing impairment who have problems with conventional air-conduction hearing aids. Patients with Down's syndrome are especially prone to these problems, due to narrow external ear canals or chronic otitis.[8] The incidence of hearing loss in children with Down's syndrome is generally reported to be as high as 78%. A combination of narrow and curved ear canals, glue ear, chronic ear infection with cholesteatoma and in some cases even (progressive) sensorineural hearing loss, contribute to the high incidence of hearing loss in children and adults with Down's syndrome.[8,9] Down's syndrome is associated with various developmental difficulties, including delayed motor and limited cognitive skills, such as speech and language acquisition and limited short-term memory abilities. In this group of patients particularly, it is essential to do

everything to maximise their hearing, so that social communication and learning abilities are not compromised even further Therefore, this specific group is expected to benefit greatly from optimal hearing aid fitting The BAHA system is a potentially significant tool in our armamentarium to improve hearing in Down's syndrome

Initially, patients with moderate mental retardation were excluded for this BAHA treatment This study showed that a BAHA produced rewarding results in such patients No extra problems occurred with the handling of their BAHA and only 3 cases (14%) mentioned problems with cleaning the percutaneous contact All 22 patients were still using their BAHA 7 days a week and for more than 8 hours a day The percentage of lost implants in this study was comparable with that reported by Priwin et al in a recent study on the BAHA (9 %) [10] A study by Reyes et al on 149 patients aged between 3 and 88 years who were fitted with a BAHA also showed a comparable rate of implant loss (8 7%) [11] Percutaneous connection can be impaired by general skin disorders, such as psoriasis or poor hygiene [12 13] In 1 case, psoriasis capitis was present at time of implantation, which may explain why the implant was lost In the other patient the reason for implant loss was unclear, but poor integration or trauma that went unnoticed by the care providers, form reasonable explanations Regular follow-up is recommended to prevent the progression of adverse skin reactions, because these can influence the success or failure of the entire system In our patient group with special needs, the patient and the care providers were made responsible for checking and taking care of the BAHA system The yearly check-ups can be used to give additional information to the care providers about how to handle the BAHA It may be worthwhile to hold even more frequent check-ups in this special group of patients If necessary, the skin around the implant can be cleaned during these visits

Remarkable improvements in daily activities in 5 of the patients showed that the quality of life issue of having better job prospects, particularly applied to our patient group Before the BAHA was fitted, some of the patients could not participate in a working situation, whereas with their BAHA, communication abilities improved to such an extent that they were able to accept a job in a special setting. The work supervisors reported that these people had been given jobs because of the positive effect of the BAHA on social communication

Audiology

In audiological terms, subjects with severe mental retardation are typically classed as difficult to test [14 15] By obtaining a wide range of data and calling upon specially trained audiologists, many gaps in information can be overcome This study was

performed on patients with moderate mental retardation and it was possible to obtain audiological information from them. During follow-up, there is a tendency for such patients to show slight progression in hearing impairment, which may distort the measurements to the detriment of the BAHA. However, the audiometric evaluations in our patient group showed clear audiometric benefit, besides better wearing comfort and obvious improvements in communications abilities. In figure 1, mean free-field thresholds with the BAHA approached the mean bone-conduction thresholds, which suggests that the BAHA is an effective treatment to compensate for the air-bone gap in this patient group. In the subgroup of 9 patients, the mean free-field thresholds with the BAHA improved by a mean of 9 dB compared to the measurements with the previous hearing aid. This clear difference was not statistically significant (P= 0,098). Speech recognition scores at 65 dB showed an equal pattern to that in the control group of BAHA users (Figure 3). Therefore, treating patients with moderate mental retardation with a BAHA system produced results that were comparable with those obtained from controls. Consequently, moderate mental retardation should no longer be considered as a contraindication. Still, careful selection procedures are essential.

It may be worth considering rehabilitation with a BAHA Softband in very young children with moderate mental retardation. This group should especially experience the benefit of an early start in rehabilitation. The early application of the BAHA Softband to very young children with syndromic bilateral aural atresia, in whom surgical intervention was not yet an option proved to be a valid treatment.[16] Therefore, the BAHA Softband is not only an option for the early treatment of young children, but also for very young children with moderate mental retardation, such as in the Grouchy syndrome.[16] In view of the potential of the BAHA system in the rehabilitation of our patient group with moderate mental retardation, the exclusion criteria for the BAHA system should be reconsidered.

Conclusions

In our patients with moderate mental retardation the BAHA was well-accepted. The extrusion rate seemed to be comparable with that of the non-mentally disabled BAHA users, especially children, who are considered to be more vulnerable. Implementation of the BAHA in patients with mental retardation was an effective hearing aid treatment that may have important positive effects on activities at school or at work. Some of these patients had been unable to reach

their full potential until they received BAHA treatment. Extending the indications for the BAHA to this special patient group is recommended.

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2.2

The bone-anchored hearing aid in patients with moderate mental retardation: impact and benefit assessment

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Abstract

Objective: To assess the impact and the subjective benefit of BAHA implementation in patients with hearing impairment combined with moderate mental retardation.

Design: Case control study using 2 validated patient oriented instruments.

Setting: Tertiary referral centre.

Patients: Twenty-two patients with moderate mental retardation and conductive or mixed hearing loss.

Method and results: BAHA implementation in patients with moderate mental retardation showed by using the Glasgow Children's benefit Inventory (GCBI) and the Listening Inventory For Education (LIFE) a subjective benefit, which was comparable with the control group and the results of earlier studies.

Conclusions: The BAHA proved beneficial in most patients with hearing impairment and moderate mental retardation. Extending the indications for BAHA application to this special patient group shows to be a very valuable option.

Introduction

Initially, in 1987 with the introduction of the BAHA patients with mental retardation were excluded from this treatment, because of doubts concerning the care provided for the percutaneous implant and surrounding skin In Nijmegen since 1996 22 patients with moderate mental retardation and conductive or mixed hearing loss have been accepted for this BAHA treatment The good clinical and audiological outcome was published recently [1] Among those patients, 4 were even fitted despite quite a limited air-bone gap Mean free-field thresholds showed a clear mean improvement of 9 dB compared with the previous hearing aid The BAHA was well accepted by these patients and was being used for most of the day Another multi-centre study on the outcome of BAHA application in patients with Down syndrome showed also a good outcome [2]

A number of recent studies have evaluated subjective results with the BAHA in patients with normal intelligence [3 7] One of these studies noted significant reductions in hearing disability and handicap by using the HHDI instrument, a patient oriented questionnaire [5] Several studies that obtained data with the Glasgow Benefit Inventory (GBI) reported overall improvement after fitting the BAHA-system [3 4 6 7]

The aim of this study is to provide subjective results of BAHA application in those 22 patients with moderate mental retardation Subjective benefit and effect on listening and learning capabilities of BAHA treatment was measured with parent and care provider directed instruments Information was obtained using 2 validated disease-specific instruments the Glasgow Children's Benefit Inventory (GCBI) and the Listening Inventory For Education (LIFE)

Methods

Patient Characteristics

Between July 1996 to June 2004, 22 consecutive patients (8 men and 14 women) with congenital moderate mental retardation and conductive or mixed hearing loss were selected for BAHA implantation at the Radboud University Medical Centre Nijmegen The majority of these patients had Down's syndrome (12 patients) Mean age at the time of implantation and at the time of completing the instrument was 36 years (range 7-73 years) and 39 years (range 10-74 years), respectively Interval between implantation of the BAHA and completing the instruments varied

from 5 months to 96 months (mean 36 months). A separate paper presented the clinical and audiological results obtained before and after BAHA implementation.[1]
In this group of 22 patients, 2 patients were receiving special education, 7 had supported employment or jobs at sheltered workshops, 9 patients were visiting a day care centre for the mentally handicapped and 3 had independent work. One patient did not have an occupational setting.
Before care providers filled out the instruments, patients had at least 8 weeks of effective experience with their BAHA, to allow time to become accustomed to the hearing aid. As the first 6 weeks after implantation were used for osseo-integration,[8] the total elapsed time between the operation and filling out the instruments was at least 14 weeks.

To be able to make clear comparisons, a control group was constructed. We paired each of the study subjects with a patient in our database with the same age and with the same implantation date. The control group comprised 22 patients with a mean age of 40 years at the time of follow-up and a mean follow-up of 35 months. There was no implant loss in this control group.
All the patients (including control group) were using their BAHA 7 days a week and for more than 8 hours a day.

Instruments
The Glasgow Children's Benefit Inventory (GCBI)
In this study, we used the children's version of the Glasgow Benefit Inventory (GBI), because of the moderate mental handicap of our patient group. The GCBI[9] was completed by one of the patient's family care providers and by one of the care providers involved in daily care at institutes. It was a parent-completed instrument, used in situations with young children, who usually lack the necessary skills in language and abstract reasoning to complete such an instrument themselves. As patients with mental retardation have as result a difficulty in completing a questionnaire, the use of an instrument, which is completed by a second person on behalf of the patient, was obligatory. The GCBI is an instrument for a retrospectively applied measure specifically worded to assess the benefit of an intervention in children and it is eminently suitable for use in paediatric otolaryngology. Outcomes e.g. benefit and improvement in the patient's state of health, can be measured and compared over a wide range of otolaryngological procedures, including fitting a BAHA.[9] The Dutch version used in this study was translated by two English translators. Their two versions were compared, adapted and the final version was agreed upon in a plenary meeting.

Possible scores on each question range from –2 to 2. A score of –2 reflects a poorer outcome, a score of 2 reflects improvement and a score of 0 reflects no change in outcome. A summary score on the GCBI was calculated by assigning a numerical value from –2 to +2 to each individual response, then adding them together, dividing by the number of questions (24) and multiplying by 50 to produce a result on a scale from –100 (maximum deterioration) to +100 (maximum improvement).

Four domains are covered by the instrument: 7 questions relate to emotional benefit, 5 questions to physical health, 4 to improvement in learning ability and 5 to vitality. For the corresponding questions see Figure 1.

In the control group the analogous adult instrument was used, the GBI, because they were able to complete the instrument themselves.[10]

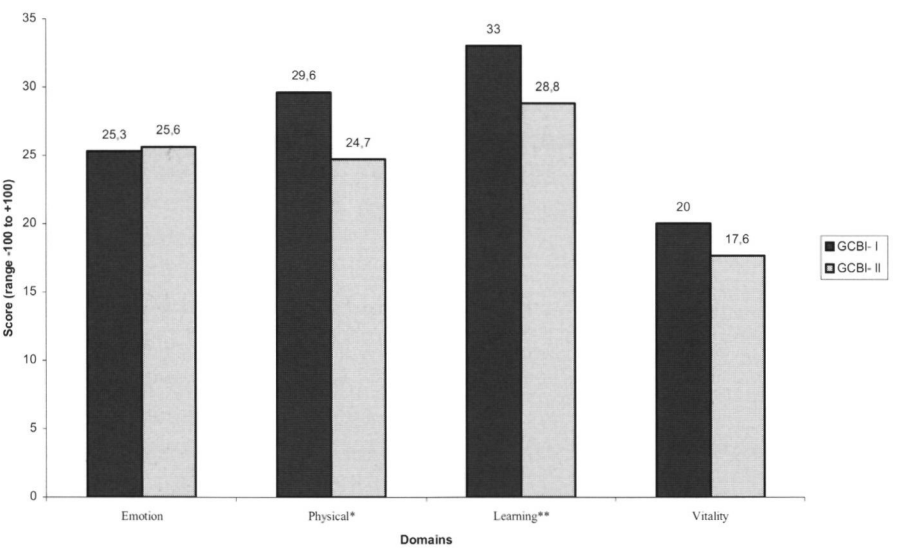

Figure 1. Results of the Glasgow Children Benefit Inventory (GCBI) per domain.
Total mean score of all patients per domain. E=emotion, corresponding questions 8,9,11,12,17,19 and 20. P=physical health, corresponding questions 1,14,22,23 and 24. L=learning, corresponding questions 4,12,13 and 15. V=vitality, corresponding questions 5,6,7,10 and 21.
* Difference not significant (p=0.050); ** Difference not significant (p=0.458).

Listening Inventory For Education (LIFE)

The second instrument used in this study was the Listening Inventory For Education (LIFE).[11] This is a valid and reliable measurement tool, in a post

intervention format, to document the effectiveness of an intervention used to improve the listening and learning capabilities, such as fitting a BAHA. Owing to the problems our patients have with reading and writing, the format of teacher appraisal was chosen and completed by the patient's care providers involved in daily care at institutes. The LIFE identifies changes that occur in attention, participation and learning.[11,12] It includes 16 questions that are related to specific areas of improvement in behaviour or learning. Respondents indicate how much they agree with the statements.

Composite scores were obtained by adding together the first 15 questions on the –2 to 2 scale. The sixteenth item was scored on a range of -5 to 5, so the maximum achievable score was 35 and the minimum achievable score was -35. A score of +15 or higher reflected that the intervention had produced positive changes and was beneficial. A score of less than 15 would be interpreted as less change or less benefit, while a score less than 4 reflected no difference. Scores of higher than 26 represent a strong positive changes and the intervention can be seen as highly beneficial.

Answers to the GCBI were given by the care provider in the daily living situation (parents, sheltered housing, home for the mentally handicapped or surrogate family home). The care provider in the daily situation (work, education, occupational therapy) was asked to fill out the GCBI and the LIFE.

To analyse the results, a two-tailed non-parametric test was used. $P< 0.05$ was chosen as the level of significance. Pearson's correlation test was used to analyse correlations. Results were computed using the SPSS software package (version 11.0; SPSS Inc, Chicago, Ill)

Results

Instruments

The response rates to the questionnaires were 80% overall, 100% to the GCBI in the daily living situation (I), 77% to the GCBI in daily working situation (II), and 73% in the control group. Response rate to the LIFE was 59%.

GCBI

Data of all 22 patients on the questionnaire in the daily living situations (GCBI-I) were complete. In 5 patients no answers could be obtained about the daily working situation (GCBI-II): 3 (nos. 10,11, 20) of them had a different daily working situation since receiving their BAHA, so their current care providers were

unable to make a good comparison, 1 patient (no. 12) did not have an occupational setting and 1 (no. 5) patient was temporarily, due to circumstances, unable to work.

The GCBI demonstrated a subjective overall patient benefit with a total mean score of +30 (n=39). Total mean scores on the GCBI (I) and GCBI (II) were +30 (n=22) and +28 (N=17), respectively. In the control group the total mean score was +32 (n=16). In the GCBI replied by the family care providers (daily living situation) no negative answers were given. The overall outcomes on the GCBI (I) and GCBI (II) showed no correlation (p= 0.228), but neither a significant difference (p= 0.859).

Viewed per question, benefit varied widely. All responses were positive regarding the overall life after BAHA fitting (question 1). Scores were above average in answer to the questions about effect on life and learning, happiness and self-consciousness (questions 2, 13, 18 and 19, respectively). Responses to progression in development and change in behaviour (questions 3 and 4) were also higher than average. Enjoyment of food, sleeping at night and improvement in self-care (washing, dressing, etc.) showed only slight changes, with scores of less than 10 (questions 6, 7 and 20, respectively). Effects on learning, question 13, were considerably higher on the GCBI (II) than on the GCBI (I) (45 versus 32). Scores on social functioning, question 9, were higher on the GCBI (I).

Separate analyses were performed on four domains (physical, vital and emotional benefit and improvement in learning) of the GCBI (I) and GCBI (II) (Figure 1). There were positive changes in physical health, with scores of +29.6 and +24.7, respectively. The improvements in learning were also remarkable (+33,0 and +28,8).

LIFE

All except for one of the patients' care providers returned the LIFE questionnaire. Eight of them were incomplete for various reasons. The most frequent reason was insufficient knowledge of the patient before the BAHA was fitted, which made it difficult to make comparisons. For three (nos. 5, 9,12) patients the questionnaire was not appropriate for their situation according to their care providers. A total of 13 questionnaires could be evaluated.

The overall mean score per question on the LIFE was +28. Strong positive changes were seen in following instructions, answering questions correctly and paying attention while listening in small groups (question 6, 7, 13 and 16, respectively). These scores were all above the mean of +28. No negative answers

were given. Only the question about video instructions did not show any difference.

The total score assigned to each patient across the 16 behaviours can be used as the criteria for success of the intervention.[11] Figure 2 shows the number of patients with significant behavioural changes as a result of using their BAHA in the daily working situation (work, education and occupational therapy). In the 13 evaluable questionnaires, 1 patient rated the BAHA as "highly successful", 6 rated it as "successful", 5 rated it as "minimally successful and 1 as "no difference". No negative changes were reported.

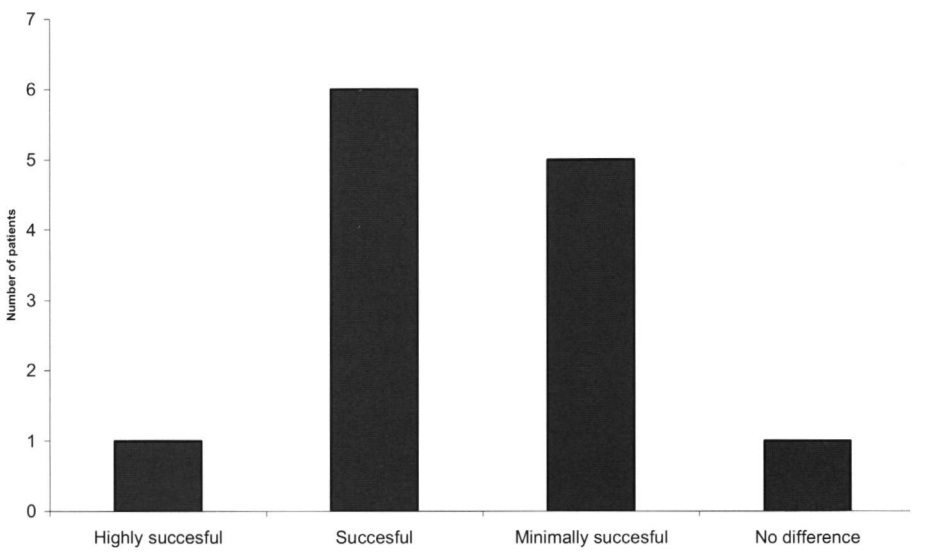

Figure 2. Results of the Listening Inventory For Education (LIFE).
Number of patients who showed significant behavioural changes as a result of using the BAHA. Highly successful (>26), successful (15-25), minimally successful (15-5) and no difference (<4).

Discussion

The aim of this study was to report on the subjective benefit of the BAHA application and its effect on listening and learning capabilities in this special group of patients. Sheehan et al reported most recently in 2006 about BAHA application in 43 patients with Down syndrome.[2] This multi centre retrospective study showed a high level of satisfaction with the BAHA amongst patients, parents and

caretakers A subjective evaluation with a health related questionnaire was not included To the best of our knowledge this is the first study that measured the benefit after BAHA implantation in patients with hearing loss combined with a moderate mental retardation The children's version of the GBI was used, because of the moderate mental handicap of the patient group The study group of 22 patients had a mean age of 39 years and a follow-up of between 5-96 months Two implants (9%) were lost in this small series of patients with moderate mental retardation (n=22) [1] The percentage of loss of the titanium implant is referred to be between 3-10% in different clinical BAHA studies [13] The outcome may be influenced by the time of follow-up The results presented in this study are in the range established in previous series with patients without a moderate mental retardation

The response rate to the questionnaires was 80% Mean overall benefit score shown by the GCBI was +30 Comparable results were observed in the control group who filled in the analogous adult instrument, the GBI This group had the same mean age and mean follow-up duration Their total mean score was +32

Retrospective studies that used the Glasgow Benefit Inventory (GBI) also reported subjective improvements after BAHA fitting In 2004, Mc Larnon et al studied 69 BAHA recipients with a mean age of 49 years [6] Patients were divided into the following subgroups discharging mastoid cavities, chronic active otitis media, congenital ear anomalies, otosclerosis and acoustic neuroma They had all been using their BAHA for at least 3 months (some for many years) Response rate to the GBI was 73% The mean score on total benefit in the entire group was +33 Total scores in each subgroup were discharging mastoid cavities +30, chronic active otitis media +37, congenital ear anomalies +45, otosclerosis +28 and acoustic neuroma +24

Dutt et al evaluated the benefit of wearing a bone-anchored hearing aid (quality of life) by means of the GBI in 2002 [3] They included paediatric and adult patients with a mean age of 9 and 45 years, respectively The study group comprised 227 patients and the response rate was 72% Follow-up varied from 6 months to 11 years Results in each of the three individual subscales were displayed as Box and Whisker plots Median scores on the general, social and physical subscales were + 40, +33 and +33, respectively Our results fell between their 25th and 75th percentiles

The study by Arunachalam et al in 2001 used the GBI to quantify changes in quality of life in 51 patients with a BAHA [4] Mean age was 45 years, the response rate was 85% and follow-up was at least 12 months Total mean benefit score was +31

Scores on the general, social and physical subscales were +34 (range 27-48), +21 (range 12-37) and +10 (range 2-26), respectively. Benefit scores on each subscale were obtained from three separate groups: congenital atresia, discharging mastoids and chronic otitis media. Values were compared to those reported by multichannel cochlear implant users. Maximum improvement was observed in the group with congenital atresia. In the study by Arunachalam et al, comparisons were made between middle ear surgery and cochlear implantation. Mean benefit scores were +17 and +40, respectively.[10] Cochlear implant users had an average total benefit score of +41 with the GBI in the study performed by Castro et al in 2005.[14] A summary of these results is shown in Figure 3. In our study group the BAHA led to comparable levels of benefit as those in other BAHA users, especially in the subgroups of chronic active otitis media and discharging mastoids, i.e. the major indications in our patients. When the values were compared to those recorded after other otological procedures, it was found that the BAHA was more beneficial than middle ear surgery in patients with discharging mastoids, but only slightly less beneficial than multichannel cochlear implantation. This was an important observation, because it supports the recommendation to extend the criteria for fitting a BAHA.

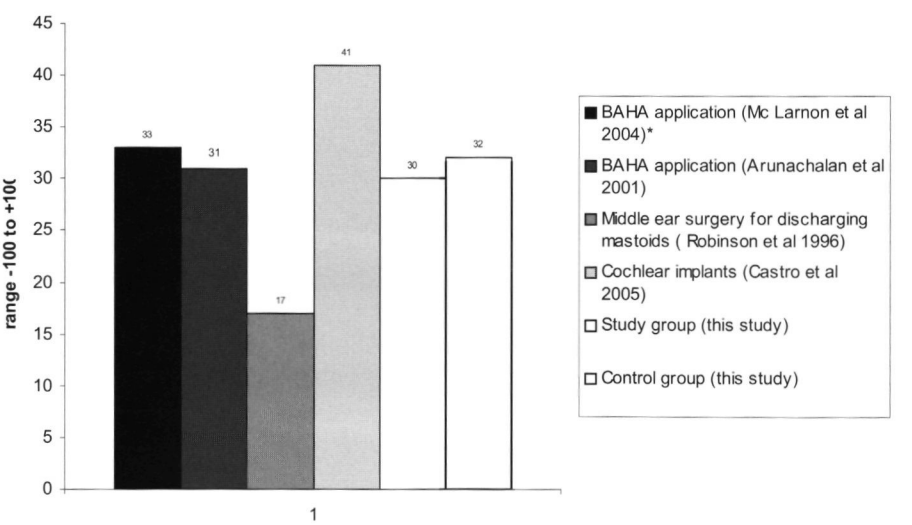

Figure 3. Benefit of different otological procedures measured with the Glasgow Benefit Inventory (GBI). Note: The GBI children version was used in the "study group".
* Average mean total scores in the subgroups chronic active otitis media and discharging mastoids (Mc Lamon et al 2004).

The Listening Inventory For Education (LIFE) can be used to identify changes in attention, classroom/ workshop participation and learning after an otological intervention, such as fitting a BAHA In this study, measurements were obtained on the daily working situation by asking the care providers to fill out the questionnaire Responders showed to be convinced about the positive effect of the BAHA in this moderate mental disabled patient group Besides the subjective measured improvements an improvement in daily activities in at least 5 patients was reported Three of them (nos 10,11, 20) were able to have a different daily working situation since receiving their BAHA Two out of these 3 patients were competent to perform independent work instead of supported employment The positive effect of the BAHA on social communication was the main reason for this change in working situation

As we compare the audiometric results of these patients to the scores on the questionnaires we have to discuss a few patients separately [1]

In 1 patient (n 7), with an air-bone gap of 9 dB and moderate scores on the GCBI and LIFE, speech recognition scores at 65 dB improved with 45% Another patient (n 8) had an air-bone gap of 14 dB and a negative score of –6 on the GCBI This patient showed to have less benefit of the BAHA Although a speech recognition score of 65 % at 65 dB with a BAHA was reached

Besides audiometric improvement after fitting a BAHA improvements can also be expected in user-comfort and in reduction of ear infections In the majority of patients, the indication for fitting a BAHA was chronic otitis and otorrhoea caused by wearing a conventional air-conduction hearing aid Application of the BAHA reduced these problems considerably, which was reflected in the score on the subscale health (Figure 1) It is likely that a large proportion of the benefit can be attributed to the improvement in physical health Moreover, the BAHA is known to be more comfortable to wear than a conventional bone-conduction hearing aid This is also reflected in the fact that all 22 patients were using their BAHA for 8 hours or more a day, 7 days a week Particularly patients with difficulties in development of social communication need good hearing rehabilitation Problems with hearing aid fitting due to recurrent ear problems can be overcome by the application of a BAHA

Conclusion

Implementation of the BAHA in patients with moderate mental retardation was beneficial and improved their listening and learning capabilities, according to their

mean scores on the GCBI and the LIFE. Comparable results were observed in the control group (GBI) and in earlier studies. Extending the indications for BAHA implementation to this special patient group seems to be a valuable option.

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3.1

BAHA system application for unilateral congenital conductive hearing impairment: audiometric results

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Abstract

Objective: To study the audiological outcome of bone anchored hearing aid (BAHA) application in patients with congenital unilateral conductive hearing impairment.

Study design: Prospective audiometric evaluation on 20 patients.

Setting: Tertial referral centre.

Patients: The experimental group comprised 20 consecutive patients with congenital unilateral conductive hearing impairment, with a mean air-bone gap of 50 dB.

Methods: Aided and unaided hearing was assessed using sound localization and speech recognition-in-noise tests.

Results: Aided hearing thresholds and aided speech reception thresholds were measured to verify the effect of the BAHA system on the hearing acuity. All patients fulfilled the criteria that the aided speech reception thresholds or the mean aided sound field thresholds were 25 dB or better in the aided situation. Most patients were still using the BAHA almost every day.
Sound localization scores varied widely in the unaided and aided situations. Many patients showed unexpectedly good unaided performance. However, non-significant improvements of 3.0° (500 Hz) and 6.9° (3000 Hz) were seen in favour of the BAHA.
Speech recognition in noise with spatially separated speech and noise sources also improved after BAHA implantation, but non-significantly.

Conclusions: Some patients with congenital unilateral conductive hearing impairment had such good directional hearing and speech-in-noise scores in the unaided situation that no overall significant improvement occurred after BAHA fitting in our set-up. Six out of the18 patients with a complete data set did not show any significant improvement at all. However, compliance with BAHA-use in this patient group was remarkably high. Observations of consistent use of the device are highly suggestive of patient benefit. Further research is recommended to get more insight into these findings.

Introduction

It is still a challenge to achieve binaural hearing in patients with congenital unilateral conductive hearing impairment For minor congenital middle ear anomalies conventional microsurgery of middle ear will be successful in about 80% of the cases [1 2] In congenital ear canal atresia only the cases with minimal involvement of the ear canal, such as in class I and IIa atresias, microsurgery of the ear may be an option to achieve binaural hearing [2] However, very few of the total number of patients with unilateral congenital ear canal atresia are suitable candidates for microsurgery Moreover surgical intervention is not usually performed before the age of 6 years in children with ear canal atresia, or before the age of 10 years in children with isolated congenital ossicular chain anomaly Wilmington et al[3] measured binaural hearing skills in patients with unilateral congenital conductive hearing loss before and after successful reconstructive surgery On a group level there were significant improvements in several tests However, the post- surgery scores were still poorer than those found in subjects with normal hearing The same tendencies were also visible in their directional hearing experiments, but remarkably, about one third of their patients had a fairly good score before surgery Apparently, these patients had learnt to compensate for their sound localization impairment Snik et al also studied patients with unilateral operable conductive hearing impairment They reported that the results were within the normal range 1 year after surgery [1] An alternative for surgery is to fit a conventional air conduction hearing aid However, in patients with complete bony ear canal atresia, this is not an option Furthermore, in cases with a large air-bone gap, very high levels of amplification with an air conduction hearing aid are needed which cannot always be provided owing to feedback problems A better solution seems to be a bone conduction hearing aid, such as the BAHA system, which is a semi-implantable percutaneous bone conduction device The BAHA system is a very promising option, as it avoids most of the drawbacks of the conventional transcutaneous bone conduction devices [4]

Application of the BAHA system to restore binaural hearing in patients with unilateral conductive hearing loss was successful realized in patients with acquired etiology [5 8] Hol et al reported significantly improved sound localization, speech recognition and subjective benefit in 18 patients with an acquired unilateral air-bone gap [7] In patients with unilateral conductive hearing impairment due to unilateral congenital (n=6) atresia an improved binaural summation and an improvement in masking level difference was seen by Chasin and Wade [8] Wazen et al reported in 2001 that in a group with mixed and conductive unilateral hearing

impairment (congenital and acquired) the speech recognition performance in the BAHA-aided condition was comparable with the patient's best score in unaided condition.[5] Directional hearing function was not measured in this study.
In an earlier pilot study, Snik et al reported in 2002 that although the BAHA effectively improved directional hearing in the patients with acquired unilateral conductive loss the results were ambiguous in two patients with unilateral congenital conductive hearing impairment.[6] Directional hearing tests did not show any improvement in these two patients after BAHA fitting, because their localization abilities were good in the unaided condition. It remained unclear why these patients had such good localization performance already. To find out whether this is a consistent finding and to gain more insight into audiological performance we needed test results from a larger group of patients. Over a period of 8 years, 20 patients with unilateral congenital conductive hearing impairment have been fitted with a BAHA. These patients were invited for audiometric evaluation that consisted of speech reception in noise and directional hearing tests.

Patients and methods

Patients

The study group comprised 20 patients with (sub)normal hearing in one ear (further referred to as the normal ear) and congenital conductive hearing impairment in the other ear. They had all received a percutaneous BAHA. Table 1 gives an overview of the patients in this study group, including their air conduction thresholds in the normal ear and impaired ear. Two of the patients participated in the previous pilot study (nos 2 and 6).[6] BAHA treatments have been performed in the period of 8 years between July 1998 and August 2005. Both adults and children were included. The age of the adults (nos 1-9) varied between 18-61 years and the children's age (nos 10-20) varied from 6 to 14 years. Mean audiometric data of all the patients are given in table 2. The conductive hearing loss was the result of unilateral congenital ear canal atresia in all cases except for 2 (nos 4 and 19), in whom it was the result of a congenital ossicular chain anomaly. (Table 1) Patient no. 4 had congenital ossicular chain anomaly with an additional complication: the facial nerve crossed the footplate. The second patient (no. 19) had stapes ankylosis and a too short long process of the incus. In this case the parents refused a stapedotomy procedure, because of the risk of evoking inner ear damage.

Table 1. Patient characteristics age at implantation, type of meatal atresia*, average pure tone audiometric measurements ($PTA_{0.5\ 1\ 2\ 4}$) at the frequencies 500, 1000, 2000 and 4000 Hz in the normal ear and impaired ear and the mean air-bone gap in the impaired ear averaged at the frequencies 500, 1000, 2000 and 4000 Hz. * Classification to Cremers et al.[2]

Patient No.	Age at implantation (years)	Meatal atresia, type	BAHA side	PTA normal ear (dB)	PTA impaired ear (dB)	Mean air-bone gap impaired ear (dB)
1	61	IIB-III	AD	9	68	50
2	40	IIB-III	AD	19	61	41
3	31	IIB-III	AD	5	76	59
4	22	No atresia	AS	13	54	30
5	23	IIB-III	AD	9	88	53
6	18	IIB-III	AS	4	69	60
7	18	IIB-III	AD	6	68	54
8	18	IIB-III	AD	4	70	49
9	20	I	AS	13	48	45
10	13	IIB-III	AD	11	73	59
11	6	IIA	AD	8	53	49
12	6	IIB	AD	6	60	50
13	10	IIB-III	AD	6	66	53
14	14	IIB-III	AD	4	55	51
15	6	IIA	AD	5	56	49
16	10	IIA	AS	15	65	49
17	5	IIB-III	AD	9	53	40
18	6	IIB-III	AD	30	89	58
19	12	No atresia	AS	8	61	40
20	6	III	AD	8	74	59

Table 2. Summary of the mean audiometric data obtained from the patient (n=20).
AC: air conduction, BC: bone conduction, PTA: mean hearing loss at 0.5, 1, 2 and 4 kHz

		AC					BC					Air-bone gap
		0.5	1	2	4	PTA	0.5	1	2	4	PTA	
Normal ear	average	11.8	9 5	9.5	7	9 4	8 3	5 5	7 5	5 3	6.6	
Impaired ear	average	69.8	67.8	64 5	58.8	65.2	14	13	21.5	13 3	15 4	49.8

Sound field measurements

Aided hearing thresholds and aided speech reception thresholds were measured to verify the effect of the BAHA system on hearing acuity, as described elsewhere.[1,6] During these measurements, the normal ear was blocked with an earplug and earmuff, which led to attenuation of approximately 40 dB.[6] BAHA fitting was only considered to be adequate when either the aided speech reception threshold or the mean aided sound field threshold (average at 0.5, 1 and 2 kHz) were 25 dB or better. All patients fulfilled these criteria.

Sound localization was tested in the horizontal plane with a measurement procedure that was essentially the same as that used by Hol et al.[7] Nine

loudspeakers were placed in a 240° circle bow (between -120 and 120 degrees) at intervals of 30 degrees. Stimuli consisted of 1 sec short bursts of 1/3 octave filtered white noise, with either a 500 or 3000 Hz centre frequency. Stimuli were presented at 65 dB SPL and four times per loudspeaker in random order. This resulted in 36 presentations per measurement condition. After each burst, the patient was asked to indicate which loudspeaker had produced the sound. The patients were not permitted to turn their head during the measurements. Per presentation, the difference in azimuth was determined between the position of the loudspeaker that had emitted the sound and the position of the loudspeaker indicated by the patient. The mean absolute error in azimuth, called MAE, per measurement condition (500 and 3000 Hz; unaided and aided) was the outcome measure. A decrease in MAE after BAHA fitting was regarded as a positive outcome.

For the children, this test procedure provided to be too lengthy. Therefore, the number of loudspeakers was reduced to 5 (60 degree intervals), which resulted in 20 presentations per measurement condition. In the original set up, the inability to localize sound and random guessing would have resulted in an MAE of 80°; in the children's version, this score was 96°.

To make intra-individual comparisons, test-retest measurements were reviewed.[6] It was concluded that a change in MAE of more than 16° in the adults could be considered as a significant change (on a 5% level). Similar data were obtained from 10 children by means of repeated measurements. A change in MAE was considered significant if it exceeded 27° and 34° in the 500 Hz and 3000 Hz measurements, respectively.

Speech reception was measured with short, everyday Dutch sentences developed by Plomp and Mimpen.[9] Speech reception thresholds (SRTs) were recorded using an adaptive tracking procedure. SRTs were measured in quiet and in noisy listening conditions. Steady state noise was used, with the same spectrum as the sentences. Speech was presented by a loudspeaker in front of the patient, while noise was presented by a loudspeaker on either the left or the right of the patient. The noise level was fixed at 65 dB. In each separate condition, the SRT was measured twice and the results were averaged. Previously the 95% confidence level for the change in the SRT was determined as ± 1.6 dB.[10]

To measure speech reception in noise in children sentence tests are not the first option, as the results prove to depend not only on hearing, but also on language competence. Therefore, words in noise are often used instead. We presented standard Dutch word lists of 11 words (monosyllables, 33 phonemes) from the front loudspeaker and continuous speech shaped noise simultaneously on the

side of the normal ear (at + or -90°). Noise was presented at 65 dB SPL, while the average speech level was 60 dB SPL. Therefore, the signal to noise ratio (S/N) was fixed at –5 dB. Phoneme scores were obtained using two lists per measurement condition, i.e. 22 words. For training purposes, one list of words was presented at an S/N of 0 dB. Differences between aided and unaided phoneme scores were tested according to Thornton and Raffin.[11] All the tests were carried out in a sound-treated, double walled room in adults and children at least 10 weeks after BAHA fitting to give the patient time to adjust to the BAHA.

Table 3. Overview of BAHA use in days per week and hours per day by all 20 patients.

Patient no.	Hours/day	Days/week	Follow-up	
			weeks	months
1	4 to 8	7	15	
2	*	*	26	
3	4 to 8	4	34	
4	>8	7	15	
5	>8	7	13	
6	>8	7	16	
7	>8	6	10	
8	>8	7	15	
9	**	**	13	
10	0	0	14	
11	4 to 8	5		68
12	4 to 8	5		37
13	>8	7		35
14	2 to 4	4		36
15	4 to 8	3		36
16	>8	7		33
17	>8	7		38
18	>8	7		24
19	>8	7		18
20	>8	7		12

* Occasionally
** Temporary implant loss

Results

On the day of the measurements the patients were asked about their BAHA use. We noted the number of days a week and the number of hours a day that the BAHA was being used. Table 3 shows the results. In the adults the mean length of

time between BAHA insertion and subsequent testing was 17 weeks (10-34 weeks). The children test procedure was done in each child after more than 12 months of BAHA use with a mean of 34 months (12-38 months).

Sound localization

Table 4 presents the results of the unaided and aided sound localization tests using the 500 Hz and 3000 Hz noise bursts (with the normal ear unblocked). The table shows that some patients had a low (thus good) MAE in the unaided situation. In the adults, a mean MAE score of 34° was calculated with the 500 Hz noise in the unaided situation, whereas 80° was expected.

Table 4. Sound localization scores with 500 and 3000 Hz noise burst, expressed in mean absolute errors (MAEs) in the unaided and BAHA situations. Patients 1-10 were measured with 9 loudspeakers at intervals of 30 degrees; patients 11-20 were measured with 6 loudspeakers at intervals of 60 degrees.

	500 Hz			3000 Hz		
Patient	Unaided	BAHA	Change	Unaided	BAHA	Change
1	29	43	-14	45	56	-11
2	6	24	-17**	12	17	-5
3	84	51	32*	86	60	26*
4	30	9	21*	16	16	0
5	36	32	4	40	32	8
6	27	33	-6	12	23	-11
7	32	19	13	27	11	16*
8	64	31	33*	103	47	56*
9	11	19	-9	24	29	-5
10	18	35	-17**	29	18	11
Mean	**34**	**30**	**5**	**39**	**31**	**8**
11	47	76	-29**	97	39	57*
12	76	68	8	63	37	26
13	86	16	70*	50	52	-3
14	68	52	16	39	63	-24
15	68	73	-5	73	78	-5
16	13	24	-11	16	34	-18
17	52	13	39*	50	13	37*
18	104	70	34*	110	112	-3
19	3	21	-18	12	6	6
20	21	48	-27**	24	45	-21
Mean	**54**	**46**	**8**	**53**	**48**	**5**

* Significant improvement, ** Significant deterioration

Significant improvement of more than 16° (p=0 05) was seen with the 500 Hz noise bursts in 3 out of the 10 adult patients (nos 3, 4 and 8) Two patients (nos 2 and10) showed significant deterioration in MAE Non of the other adult patients showed any statistically significant changes A similar pattern was seen with the 3000 Hz noise bursts in the adults Their mean unaided MAE score were 43° Two out of the 10 patients (nos 3 and 8) showed improvement of more than 16° Sound localization abilities with the 3000 Hz noise did not deteriorate significantly in any of the patients On average, MAE scores improved by 4° with the BAHA in the 500 Hz noise and by 8° in the 3000 Hz noise

In the modified set-up used in the children, the MAE score due to random guessing, was calculated as 96° However, such a high value was seldom found The mean unaided MAE score was 54° On an individual level, an increment of more than 27° was a significant change in MAE in the 500 Hz condition Three out of the 10 children (nos 13, 17 and 18) showed significant improvement in this condition, whereas 2 children (nos 11 and 20) showed significant deterioration In the 3000 Hz condition, an improvement of 34 degrees or more was significant Two children (nos 11 and 17) showed significant improvement Significant deterioration was not observed

Speech recognition

Table 5 shows the speech reception thresholds (SRTs) of the adults in quiet SRTs with noise, presented on the normal side and on the impaired side, are shown as speech-to-noise ratios (S/N ratios) A decrease in SRT reflected an improvement in speech recognition and was thus defined as a positive outcome One dB decrease in S/N ratio results in 15% better speech recognition [9] The unaided minus BAHA condition is denoted as change Therefore, an improvement in speech recognition is always denoted as a positive change

Use of the BAHA had a significant effect (improvement of >1 6 dB) on speech recognition in quiet in 4 out of the 10 patients (nos 3, 5, 6, and 9) When the speech was presented in front of the patient and the noise on the normal side, 4 patients (nos 4, 6, 7 and 9) showed improvement of more than 1 6 dB On average, the S/N ratio increased by 1 1 dB, which was not significant (p<0 05)

Generally, patients with unilateral hearing impairment do not usually experience much hindrance in the unaided situation when their impaired side is exposed to the noise However, after the BAHA has been fitted to that side, the amplified noise might be bothersome Therefore, if a patient has no trouble understanding speech in the aided situation in spite of hearing amplified noise on the BAHA side, then the score is expected to be around zero as shown in our previous study [7] In

this listening condition, we found fairly wide inter-individual variation. One patient (no. 10) showed significant deterioration of > 1.6 dB, whereas one patient (no. 8) showed an unexpected significant improvement of > 1.6 dB. On average, the change in S/N ratio with the BAHA was 0.4 dB, which was not statistically significantly different from zero.

Table 5. Speech reception thresholds (SRTs) in quiet in the adult patients (n=10) in the unaided and BAHA situations (unaided minus BAHA is denoted as change).

	Speech in front, without noise SRT results (dB)			S/N results, speech in front Near normal ear S/N (dB)			Position of the noise source Near BAHA (impaired) ear S/N (dB)		
Patient no.	Unaided	BAHA	Change	Unaided	BAHA	Change	Unaided	BAHA	change
1	28.0	29 2	-1 2	-0.4	0 4	-0.8	-4 1	-4.0	-0 1
2	38 1	37.8	0.3	-1.1	-1.1	0.0	-3 0	-2.6	-0.4
3	31.7	28 5	3.2*	-0.7	0 3	-1.0	-3 9	-4.6	0.7
4	32.7	34 2	-1.5	0 6	-2 0	2.6*	-4 0	-4.1	0 1
5	33.7	30 5	3.2*	-0.6	0 3	-0.9	-4.0	-5.0	1 0
6	26.5	20 6	5.9*	-3.0	-7 4	4 4*	-10.8	-11 2	0 4
7	21.8	21.6	0.2	-1.7	-4 0	2 3*	-6.8	-7.4	0.6
8	25.8	25 8	0.0	-6.6	-7 4	0 8	-6.1	-11 2	5 1*
9	37.1	33.4	3.7*	3.0	-0 2	3 2*	-1.0	-1.0	0.0
10	35.6	36.2	-0.6	-1.4	-1.6	0 2	-5 8	-2.3	-3.5**
Mean	31.1	29.8	1.3	-1.2	-2.3	1.1	-5.0	-5.3	0.4

* Significant improvement; ** Significant deterioration

Table 6 shows the speech reception results of the group of children (nos 11-20) tested with words in two conditions (unaided and aided with the BAHA) in a speech-to-noise ratio (S/N ratio) of –5 dB. In 2 of the children, no measurements were available in the unaided condition. Significant changes between the unaided and aided conditions are indicated; significant improvements were found in 5out of the 8 children with a complete data set. The overall results showed that the BAHA led to 23% more of the phonemes being repeated correctly.

To study the relation between test outcomes and BAHA use we correlated BAHA use with an individually determined summary score based on the number of subtests with a statistically significant change after BAHA fitting. This summary score comprised the three subtest scores on the 500 Hz directional hearing test, the 3000 Hz directional hearing test and the speech in noise test with the noise presented near to the normal ear. In this way, summary scores could be obtained from 18 subjects (except for nos 16 and 19 owing to absent unaided speech in noise data). Hypothetically, the summary score could range between –3 and +3.

One other patient had to be excluded, owing to recent loss of the implant (patient no 9). The Spearman rank correlation coefficient was 0.55 (n=17, p=0.02), thus statistically significant. This significant positive correlation between BAHA use and outcomes validated the measurements.

Table 6. Speech reception results of patients 11-20 (children) expressed as the percentage of correctly repeated phonemes in a speech to noise ratio (S/N ration) of – 5 dB Words were presented in front of the patient at 60 dB, while the noise (65 dB) was presented by a loudspeaker on the side of the normal ear.

Patient no.	Unaided (%)	BAHA (%)
11	30	55*
12	61	67
13	61	64
14	52	64
15	67	88*
16	-	76
17	55	94*
18	33	67*
19	-	65
20	45	*76
Mean (n=10)		73
Mean (n=8)	51	74

* Significant improvement according to Thornton and Raffin [11]

Discussion

Means to achieve binaural hearing in most of the patients with a unilateral congenital conductive hearing impairment as result of an unilateral ear canal atresia were still lacking. As a result of widening of the indications for the BAHA-application it was studied whether the BAHA system is a valuable option for these patients. For acquired unilateral conductive hearing impairment, first reports on a successful outcome of the BAHA application have been published. The audiometric and subjective patients' results showed improved binaural hearing.[6,7] Fitting a BAHA had a complementary effect on hearing and an obvious benefit in daily life, which provides encouragement to continue to apply the BAHA to rehabilitate patients with unilateral conductive hearing loss.[7]

In the present study, we evaluated a group of 20 subjects with congenital unilateral hearing impairment, whose mean air bone gap was 50 dB. Sound localization tests after BAHA application showed limited benefit. On a group level, the mean MAE scores in the unaided situation were 34^0 in the adults and 54^0 in

the children with the 500 Hz stimulus. When the 3000 Hz stimulus was presented, the MAEs were 43° and 53°, respectively. These scores were better than the expected random-guessing levels of 96^0 and 80^0 respectively. In the aided situation, the mean MAE values improved by 8^0 or less at the 2 test frequencies and in the 2 subgroups. In the earlier study by Hol et al in 2005, 13 patients with acquired conductive hearing loss were evaluated with exactly the same measurement set-up. The average improvement in sound localization was higher after BAHA application: viz. around 20^0. It can be concluded that in contradiction to the group with an acquired unilateral air bone gap, the patients with congenital onset had limited benefit.[7] This is the result of already unexpected good results in the unaided situation. Nevertheless, in 2 adult cases with poor unaided MAE scores (nos 3 and 8), highly significant improvement was observed with the BAHA. So far, it is not clear which patients will benefit from BAHA application in terms of sound localization. No consistent trend was visible after relating the changes in sound localization abilities to age at implantation, type of ear anomaly or duration of device use. The poor sound localization results were in agreement with the recent findings published by Priwin et al.[12] Their sound localization test set-up was largely similar to ours. They concluded that the sound localization ability of their six children with congenital unilateral conductive hearing impairment was even somewhat poorer with the BAHA than unaided.

In the group of patients with acquired unilateral conductive hearing loss (Hol et al)[7] an average improvement in SRT in quiet listening condition of 2.2 dB was found. In the present study a lower mean improvement in SRT of 1.3 dB was found (adult group). When noise was presented on the side of the normal ear, the average improvement in the S/N ratio was for the acquired versus the congenital group, 3.1 dB vs. 1.1 dB. It should be noted that the 1.1 dB benefit is low and is comparable with the effect of lifting the head shadow in patients with complete unilateral inner ear deafness who are using a CROS hearing aid.[13] When the noise was presented on the side of the impaired ear, the average change in S/N ratio was comparable between the acquired group and the congenital group. This was in agreement with a previous study in which patients with unilateral inner ear deafness had much poorer scores in this condition.[13]

The speech in noise tests with the 10 children (Table 6) showed in 5 out of 8 children with a complete data set, a significant improvement with the BAHA. This illustrates the beneficial effect of the BAHA in a noisy environment. The overall improvement was 23%.

In conclusion, the present patient group with congenital unilateral conductive hearing loss demonstrated substantially less benefit from BAHA application than

the group of patients with the acquired form This is the result of already unexpected good results in the unaided situation compared to the group with an acquired etiology For the moment a full explanation for the relatively good results in the unaided situation with directional hearing and speech understanding in noise tests in the congenital group compared to those with acquired etiology is lacking and requires additional research

However, compliance with BAHA-use in this patient group was remarkably high Observations of consistent use of the device are highly suggestive of patient benefit It can be suggested that patient satisfaction with BAHA use for monaural impairment may relate to perceptual domains (e g sound-field expansion and loudness growth effects) not tested herein and will require further study Furthermore, before clinical application, a trial period with a BAHA-headband is part of our selection procedure and might help to evaluate whether a patient is a suitable candidate for BAHA-system application for an unilateral large conduction hearing impairment

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3.2

Subjective benefit after BAHA system application in patients with congenital unilateral conductive hearing impairment

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Abstract

Objective: To study whether unilateral bone anchored hearing aid (BAHA) fitting led to subjective hearing benefit in patients with congenital unilateral conductive hearing impairment.

Study design: Prospective evaluation on 20 patients.

Setting: Tertiary referral centre.

Patients: Ten adults and 10 children with congenital unilateral conductive hearing impairment, with a mean air-bone gap of 50 dB were included.

Methods: Subjective bilateral hearing benefit after BAHA fitting was measured using two disability-specific questionnaires: Chung and Stephens and the Speech Spatial and Qualities of hearing profile (SSQ). (Children's version in the patients aged < 18 years) The Glasgow children's benefit inventory (GCBI) was also used to measure patient's health benefit after BAHA fitting.

Results: Chung and Stephens' questionnaire showed an overall preference for the BAHA in several specific hearing situations. The GCBI demonstrated an overall mean improvement of +34, which was the most prominent in the learning domain. The 10 adults showed an already good score on the SSQ in the unaided situation.

Conclusions: The BAHA was well accepted by most of the patients with congenital unilateral conductive hearing impairment. A preoperative trial of the BAHA-system with the BAHA on a headband is part of the preoperative procedure. In children with unilateral conductive hearing loss, with regard to possible childs'development and communication difficulties, intervention with BAHA can be considered as an option.

Introduction

In the past, unilateral hearing impairment was not thought to have any significant consequences because speech and language presumably developed appropriately with the aid of only one ear with normal hearing However, several previous studies[1-4] evaluated, in a review study by Lieu in 2004[5], problems with language development and performance at school of children with unilateral hearing and concluded that such children had a significantly higher risk for developmental delays than their peers with normal hearing [5] This suggests that intervention is also advisable for children with unilateral hearing loss, however, studies on hearing aid compliance in such children showed poor device use (e g Davis et al , 2002) [6] In their position statement, the Pediatric Workgroup (Bess et al , 2000) stated that in case of unilateral hearing loss amplification should be considered on a case-by-case basis, with regard to the child's development and communication needs [7]

When binaural hearing cannot be achieved by microsurgery or a conventional hearing aid in patients with congenital unilateral conductive hearing impairment because of ear canal atresia there is still the option of fitting a bone conduction hearing aid (e g the BAHA system) [8] In patients with bilateral conductive hearing loss, BAHA application proved to be an effective means to restore binaural hearing [9 11]

Quality of life aspects after medical interventions are receiving more and more attention In a study by Arunachalam et al in 2001 to quantify changes in quality of life following BAHA fitting, the Glasgow Benefit Inventory demonstrated clear improvements in the subgroup of patients with bilateral congenital atresia (n=12) [12] In 2004, Mc Larnon et al found that their subgroup of patients with congenital ear disorders (n=10) experienced the most benefit from BAHA application [13]

The promising audiometric and subjective results of the BAHA system in patients with bilateral conductive hearing loss encouraged extending the indications for BAHA application

In 1998, we started to use the BAHA system to treat patients with severe acquired or congenital unilateral conductive hearing impairment who did not have any other treatment option Audiometric and subjective assessments on a prospective group of 18 consecutive patients with unilateral acquired conductive hearing loss showed restoring of binaural hearing and high patient satisfaction [14 15] In the present study we made assessments of the subjective outcomes of BAHA fitting in

20 patients with unilateral congenital conductive hearing impairment. All had normal bilateral inner ear hearing levels and a mean air bone gap of 50 dB in the affected ear. Nine patients were older than 18 years at time of implantation, while the other 11 patients were younger than 18 years.

Methods

Patients

The study group comprised 20 consecutive patients with (sub)normal hearing in one ear (further referred to as the normal ear) and congenital unilateral conductive hearing impairment in the other ear. Implantations were performed in an 8 year period between July 1998 and August 2005. All the patients had normal bilateral cochlear function, with mean bone-conduction thresholds of 7 dB HL in the normal ear and 15 dB HL in the impaired ear, averaged over the frequencies 500, 1000, 2000 and 4000 Hz. The air-bone gap was the result of congenital unilateral ear canal atresia (n=18) or an isolated congenital ossicular chain anomaly (n=2). Congenital ear canal atresia was classified according to Cremers et al.[16] The mean air bone gap in the impaired ear was 50 dB, averaged over the frequencies 500, 1000, 2000 and 4000 Hz. Table 1 gives an overview of all the patients in our study group.

Eleven patients underwent one-stage surgical procedure, while 9 patients underwent a two-stage procedure. In one patient, implant loss did occur in 4 months after surgery due to skin infection.

Measurement instruments

Opinions about the BAHA were obtained from all adult patients (age >18 years at the time of evaluation, n=10) using the Dutch version of a disability-specific questionnaire introduced by Chung and Stephens (section A).[17] This questionnaire is one of the few that has been developed to gather patients' opinions about monaural versus binaural hearing. In order to avoid enthusiasm bias, the questionnaire was filled out when the patients had at least 6 months of experience with the BAHA. The options for answering were "with the BAHA" (i.e. binaural hearing), "no BAHA" (i.e. monaural hearing) or "no preference". Questions were also asked about (daily) use of the BAHA system and satisfaction with the BAHA.

Table 1 Patient characteristics age at the time of implantation, meatal atresia*, auricle anomaly, middle ear anomaly, BAHA side and mean air bone-gap in the impaired ear averaged at the frequencies 500, 1000, 2000 and 4000 Hz * Classification according to Cremers et al [16]

PTA, pure-tone average

Study no	Age at implantation (yr)	Meatal atresia, type	BAHA side	PTA normal ear (dB)	PTA impaired ear (dB)	Mean air-bone gap impaired ear (dB)
1	61	IIB-III	AD	9	68	50
2	40	IIB-III	AD	19	61	41
3	31	IIB-III	AD	5	76	59
4	22	No atresia	AS	13	54	30
5	23	IIB-III	AD	9	88	53
6	18	IIB-III	AS	4	69	60
7	18	IIB-III	AD	6	68	54
8	18	IIB-III	AD	4	70	49
9	20	I	AS	13	48	45
10	13	IIB-III	AD	11	73	59
11	6	IIA	AD	8	53	49
12	6	IIB	AD	6	60	50
13	10	IIB-III	AD	6	66	53
14	14	IIB-III	AD	4	55	51
15	6	IIA	AD	5	56	49
16	10	IIA	AS	15	65	49
17	5	IIB - III	AD	9	53	40
18	6	IIB-III	AD	30	89	58
19	12	No atresia	AS	8	61	40
20	6	III	AD	8	74	59

The younger patients (n= 10) and their parents/ care providers were asked to fill out the Glasgow Children's Benefit Inventory (GCBI), which is a parent-completed instrument that measures patient's benefit and improvement to the patient's state of health It is retrospective and suitable to measure benefit and improvement after an otological intervention in children Questions addressed improvements in different items (i e effect on quality of life, self-consciousness, learning, etc), scores could range from –2 to 2 A summary score on the GCBI was calculated to produce a result on a scale from –100 (maximum deterioration) to +100 (maximum improvement) The instrument covered four domains emotional benefit, physical health, improvements in learning ability and vitality

The Speech, Spatial and Qualities of hearing scale (SSQ) was used to assess benefit in spatial hearing and speech reception These are presumed to be of importance to binaural hearing As we were primarily interested in spatial hearing,

we decided to use the spatial hearing domain of the questionnaire, which consists of 16 items. (i.e. locate speaker round a table, locate dog barking, judge distance of a vehicle, etc) Patients rated themselves on each item with a score out of 10; higher scores reflected greater ability.

The recently developed children's version of the SSQ was used in 10 patients. It was based on the original version developed by Gatehouse and Noble, adapted by Karyn Galvin (The Bionic Ear Institute, Australia) and translated into Dutch by Liesbeth Royackers (Labo Exp. ORL, K.U.Leuven, Belgium). Language and situations were adapted according to how a child perceives the environment. The questionnaire contained three aspects of hearing: speech reception, spatial hearing and quality of hearing. Children completed this questionnaire under the supervision of an adult. This questionnaire has recently been developed and has not yet been validated. Answers could be given on a scale from 0 to 10. Questionnaires were filled in at least 6 months after surgery. Audiological data, obtained in this patient group, have been published elsewhere.

Analysis

The results were computed using the SPSS package (version 12). The Wilcoxon signed-ranks test and Mann-Whitney test were used. A p value of < 0.05 was chosen as the level of significance, with $p < 0.025$ in the case of a two-tailed t-test.

Results

Patient outcome measures

Adults (n=10)

Nine out of the ten adult patients returned the questionnaire of Chung and Stephens with assigned ratings and comments. One patient was waiting for re-implantation of the titanium implant. The first two questions on the disability-specific questionnaire introduced by Chung and Stephens concerned subjective satisfaction with the BAHA. Six of the patients were very satisfied, while 2 patients were satisfied. In answer to the questions about (daily) use of the BAHA, 2 patients were using their BAHA "always", which meant seven days a week for at least 12 hours a day; three patients were using the BAHA every day for 8-12 hours a day; two other patients were using the BAHA regularly, one of them for 4-8 hours a day and the other "occasionally". In the latter case, the patient only used the BAHA after a cold or during a meeting. Nevertheless, she was very satisfied with it and it gave her confidence in tricky situations such as in traffic. One patient

(no. 10) experienced less benefit in his puberty and therefore recently stopped using the BAHA after a period of daily-use. BAHA use data are presented in Table 2.

Table 2 BAHA use in days per week and hours per day by all 20 patients.

Patient no.	(h/d)	(d/wk)
1	4 to 8	7
2	*	*
3	4 to 8	4
4	>8	7
5	>8	7
6	>8	7
7	>8	6
8	>8	7
9	**	**
10	0	0
11	4 to 8	5
12	4 to 8	5
13	>8	7
14	2 to 4	4
15	4 to 8	3
16	>8	7
17	>8	7
18	>8	7
19	>8	7
20	>8	7

* Occasional user
** Temporary implant loss

According to the answers presented in Table 3, most of the patients gave preference to using the BAHA system in several every-day situations. When they were asked whether they would recommend the BAHA to another person with same hearing disability, all the patients gave a positive response.

Table 3 Answers given to questions 5 to 12 in the Chung and Stephens' questionnaire

	BAHA	No BAHA	No preference
When you are listening to speech in quiet situations do you find listening easier using	7	2*	0
When you are listening to the TV or radio, do you find listening easier using	7	1*	1
When you are listening to a conversation from a distance (of 6 m), do you find listening easier using	8	0	1
When you are listening to speech in noisy situations, do you find listening easier using	6	1*	1
When you are at a meeting, church or theatre, do you find listening easier using	8	0	0
When you have to locate sounds, e g car horn, do you find listening easier using	6	1*	2**
When you are listening, do you find it more comfortable (more relaxed and easier) using	7	2*	0
	Yes	No	Do not know
Would you recommend the BAHA to someone with the same hearing disability?	9	0	0

* No 2 occasional user
** Nos 1 and 3

The SSQ was returned by 6 out of the 9 adult patients with assigned ratings and comments The mean score in the unaided situation in the spatial domain was 4 5 on a scale from 0 to 10 Mean score with the BAHA increased to 6 8 (p=0,046) Results are presented in Table 4

Table 4 Adults' SSQ results, mean values and standard deviations on the spatial domain of the questionnaire
Literature data from an earlier study on acquired unilateral conductive hearing loss (Esch et al [21]) and a control group of 50 patients with asymmetrical sensorineural hearing loss Data inside parenteses are standard deviations

SSQ	Unaided	Aided
Present study	4 5 (2 4)	6 8 (1 2)
van Esch et al [21]	3 5 (0 6)	7 1 (0 9)
Gatehouse and Noble[22]	4 8	n a

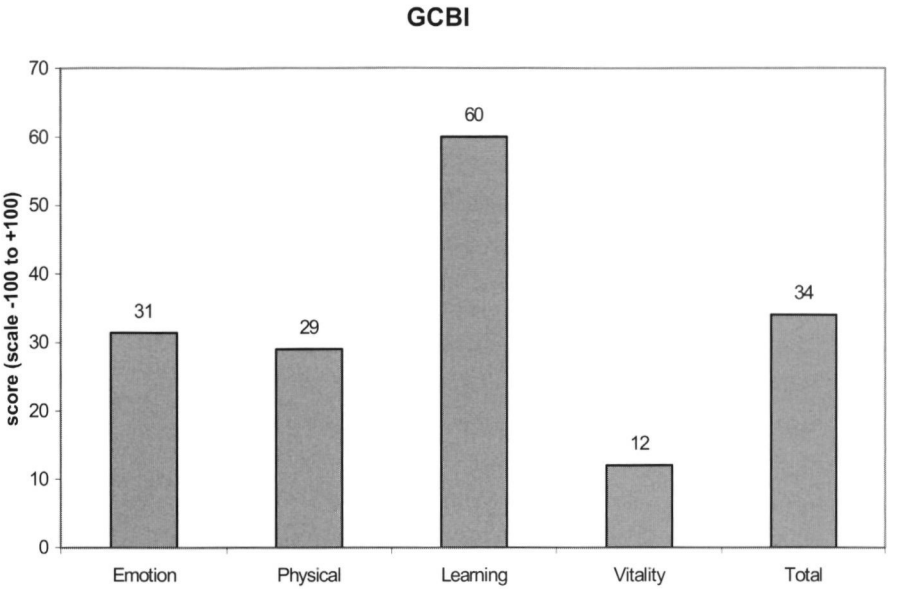

Figure 1. Results of the Glasgow Children Benefit Inventory (GCBI) per domain. Total mean score of the patients (n=10) and total mean score per domain: emotion, physical health, learning and vitality

Children (n=10)

The Glasgow Children's Benefit Inventory (GCBI) demonstrated a subjective overall benefit of + 34 (n=10). Viewed per domain, learning showed the most positive change, with a mean score of +60. The emotion domain, physical domain and vitality domain scored +31, +29 and +12, respectively (Figure 1).

The children version of the SSQ in this group (n=10) showed a total mean score of 6.6 with the BAHA, on a scale from 0 to10. Mean scores on the domains: speech, spatial and quality of hearing were 6.1, 5.6 and 7.1, respectively. Results can be seen in Table 5.

Table 5. SSQ results in the children. Mean values and standard deviations on the three domains: speech perception, spatial hearing and quality of hearing on a scale from 0 to 10.

Domain of children's SSQ	Mean	Standard deviation
Speech	6.1	2.3
Spatial	5.6	2.5
Quality	7.1	1.1

Discussion

In patients with bilateral congenital ear canal atresia, treatment with a hearing device within 6 months after birth is considered to be essential to avoid delays in speech or language development, especially now that the BAHA softband is available.[18] Children who receive early hearing intervention have significantly higher levels of receptive and expressive language, personal-social development, expressive and receptive vocabulary and vowel production.[19]

Immediate intervention is still not considered to be urgent in children with unilateral ear canal atresia, provided that the child has normal hearing in the other ear. It is rarely recommended to fit a conventional air conduction or bone conduction hearing aid, because experience has shown poor acceptance by most children. Nevertheless when speech and language development are delayed, treatment with a hearing aid, should be considered.[6,7]

The BAHA softband has proven to be effective in young children with bilateral congenital atresia, for children too young for implantation of the fixture in the skull-bone.[18] The BAHA softband might also be an option in unilateral cases if the development of the child gives reasons for concern. During early childhood, it is therefore of value to document the speech and language development in children with a severe unilateral conductive hearing loss. Our own experience with the BAHA softband has shown that a trial by children with unilateral hearing impairment when they are under the age of 6 years may help to solve any delays or behavioural problems and enable them to realize the benefit of permanent percutaneous application of a BAHA system.

Teenagers and adults with severe unilateral conductive hearing loss due to unilateral atresia often find the consequences to be a significant handicap in social settings or at work and they are more willing to try a new type of intervention to achieve binaural hearing. The adult patients are capable of pointing out the drawbacks of unilateral hearing impairment in more and more demanding environments. Although the group of adults in this study has learned to cope with their hearing loss over the years, they expressed subjective benefit after BAHA fitting.

The measurement instruments used in the adults and children in this study showed patient satisfaction with the BAHA. Most of the patients were using their BAHA nearly all day, every day of the week (Table 2). One patient (no. 10) was not convinced about the benefit of using the BAHA, so he decided to stop using it after a period of more than 12 months. Although another patient was an occasional user, she was satisfied with her BAHA and experienced considerable

benefit from using it in specific listening conditions Our adults expressed clear overall preference for the BAHA in several specific listening situations, such as in conversations, at meetings and at the theatre Except for the occasional user, none of our patients preferred "no BAHA" in any of the listening situations addressed in this study

The children's version of the GCBI used in the present study showed patient satisfaction and great improvement in learning abilities (+60) A score of +34 reflected the overall subjective benefit The latter score was similar to that obtained from a group in an earlier study (+32) in which 22 patients with bilateral conductive hearing loss and moderate mental retardation were evaluated with the GCBI after BAHA fitting In this group the overall subjective benefit score was +32 [20]

According to the spatial part of the SSQ, our adult patients experienced subjective benefit in directional hearing after BAHA fitting Scores obtained from the adults on the spatial hearing domain of the SSQ showed an overall improvement of 2 3 after BAHA fitting In a previous study by Esch et al the SSQ questionnaire was used to evaluate 8 patients with unilateral conductive hearing impairment who had received a BAHA Their deafness was due to a chronic draining ear that was resistant to medical therapy [21] Evaluation with those patients showed an overall aided score of 7 1 This was almost similar to the aided score of 6 8 in this study group

Although, the unaided score in our study group was 4 5, which was higher than the unaided score in the acquired conductive hearing impairment group [21] The unaided score of those 8 patients in spatial domain was 3 8

Noble and Gatehouse obtained an unaided score of 4 8 from 50 patients with asymmetrical sensorineural hearing loss who were fitted unilaterally [22]

The aided score for the children's group was 5 6 in the spatial hearing domain No reference data are available in the literature on the new children's version of the SSQ questionnaire Nevertheless it is reasonable to assume that these scores represent valid scores from the children in our group Priwin et al (2006) published recently data on 6 children with unilateral congenital ear canal atresia Four of them had received a BAHA system and 2 a conventional hearing aid They showed significantly improved scores in the speech recognition in noise tests (S/N ratio of 0 dB) In the less difficult test situations (S/N ratio +4 and +6 dB), there were no significant improvements compared to the unaided situation These 6 children were mainly using their device at school on a schedule that varied from rarely to frequently After device fitting, they were all well-satisfied and

experienced high quality of life. Our results confirm the subjective benefit reported by Priwin et al[23], in a larger group of patients.

In conclusion, most of our patients with congenital unilateral conductive hearing impairment showed subjective benefit after BAHA fitting. Before the decision is made to implant a BAHA system, we consider it as important for adults and children to have a trial period with a BAHA: in adults using a spring headband and in children using a BAHA softband. In this way, it will be possible to test whether expectations of a patient with unilateral congenital conductive hearing impairment correspond with the presumed effect of a BAHA.

In children with unilateral conductive hearing loss, with regard to possible childs' development and communication difficulties, intervention with BAHA can be considered as an option.

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4.1

Bone-anchored hearing aid in patients with acquired and congenital unilateral inner ear deafness (BAHA CROS): clinical evaluation of 56 cases

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Abstract

Objective: Evaluation of audiological and subjective benefit of the bone-anchored hearing aid (BAHA) as a device for, transcranial routing of sound (BAHA CROS) in 56 patients with unilateral inner ear deafness.

Study design: Prospective clinical follow-up study

Setting: Tertiary referral centre

Patients: Previous results of 29 patients were completed with a second series of 30 patients with unilateral inner ear deafness. Twenty-eight patients with acquired origin and 2 patients with congenital origin; 3 patients dropped-out during the evaluation. Therefore a total 56 patients were evaluated.

Intervention: Audiometric measurements were taken before and after BAHA CROS fitting. Subjective benefit was quantified with 4 patient questionnaires: the Abbreviated Profile of Hearing Aid Benefit (APHAB), the Glasgow Hearing Aid Benefit Profile (GHABP), the International Outcome Inventory for Hearing Aids (IOI-HA) and the Single Sided Deafness (SSD) questionnaire.

Results: Sound localization results in a well-structured test setting were no different from chance level. Patients reported some improvement in their capacity to localize sounds with the BAHA CROS in daily life. The main effect of the BAHA CROS was to lift the head shadow effect during the speech in noise test. Mean day-to-day use of the BAHA CROS in the total group was 84%. All the instruments used to quantify subjective benefit with the BAHA CROS showed positive results.

Conclusions: Poor sound localization in this larger series of patients confirms the findings of previous studies. Improvements in the speech in noise scores corroborated the efficacy of the BAHA CROS to lift the head shadow. The 4 different patient questionnaires revealed subjective benefit and satisfaction in various domains.

Introduction

Application of the bone-anchored hearing aid (BAHA) as a device for transcranial contralateral routing of sound (CROS) device in unilateral inner ear deafness has been studied at several centres [1 6] Overall, these studies reflect the efficacy of the BAHA CROS in lifting the head shadow, which is advantageous in specific listening situations, for example at the dinner table, while driving a car, or at a meeting with the speaker on the hearing impaired side Patients with unilateral inner ear deafness have only one functioning cochlea, which precludes the use of the interaural time and intensity differences that are essential for directional hearing It has been shown that application of a conventional CROS or the BAHA CROS cannot restore binaural hearing This is corroborated by reported sound localization results, which do not differ from chance level [2 4 6]

Speech discrimination in noise showed improvements with the BAHA CROS only when the speech was presented on the impaired side The patient questionnaires showed substantial subjective benefit and indicated considerable satisfaction [1 7]

To obtain more data on the BAHA CROS application, we extended our group of unilateral inner ear deafness patients from 29 to 56 individuals This offers the opportunity to evaluate separate subgroups in greater detail The test protocol comprised localization measurements, speech reception in noise tests and subjective quantification with the 4 different questionnaires, used by Hol et al [6] the disability-specific Abbreviated Profile of Hearing Aid Benefit (APHAB) to evaluate hearing aid benefit in three different communication domains and one listening comfort domain[8], the Glasgow Hearing Aid Benefit Profile (GHABP) to evaluate hearing disability, handicap, hearing aid use and benefit, residual disability and patient satisfaction with hearing aids[9], the international outcome inventory for hearing aids (IOI-HA), used internationally to assess the benefit of hearing aid fitting[10], the Single Sided Deafness (SSD) questionnaire that focuses on improvements in quality of life in patients with unilateral inner ear deafness and a BAHA [3]

Patients and methods

Patients

In this study, we completely evaluated 56 patients The data of Hol et al[6] of 29 patients were completed with a second series of 30 successive patients with unilateral inner ear deafness selected for BAHA CROS implantation Pre-

operatively, all the patients had tried a BAHA CROS on a headband for at least 1 or 2 weeks. BAHA surgery took place between November 2003 and September 2005. Mean age at the time of implantation was 48 years (range 16-71 years). During our evaluation procedure, 3 patients dropped-out: 1 patient lost the titanium implant three months after surgery and was awaiting reimplantation; 1 patient preferred a conventional hearing aid in the best hearing ear due to deterioration on that side and 1 patient experienced insufficient benefit and stopped using the BAHA.

Thus, 27 patients participated in this study: 25 with acquired unilateral inner ear deafness (after acoustic neuroma surgery n=10, trauma n=5, cholesteatoma surgery n=3, stapedotomy surgery n=1 and e causa ignota n=6) and 2 patients with congenital unilateral inner ear deafness. Mean duration of unilateral inner ear deafness was 16 years (range 1–60 years). Data on the 29 patients evaluated by Hol et al.[6] were added to extend the population to a total of 56 patients.

Hearing on the contralateral side was (nearly) normal in most of the cases: mean air-conduction threshold was 15 dB HL averaged at 0.5, 1, and 2 kHz and the bone-conduction threshold was 13 dB HL. Patient no. 49 had a significant air-bone gap in the best ear: mean air-conduction threshold was 38 dB HL and mean bone-conduction threshold was 15 dB HL. Patient no. 55 had average air and bone-conduction thresholds of 42 dB HL and 32 dB HL, respectively. Seventeen patients used a BAHA Compact, while the other 10 patients used the BAHA Classic. Table 1 presents an overview of age, gender, etiology and duration of unilateral inner ear deafness at time of BAHA surgery.

Methods

The BAHA CROS was fitted six to eight weeks after implantation surgery. This period complies with the recommended recovery period of six weeks for osseo-integration.[11] Audiometric evaluations were carried out in all the patients in the unaided situation and after at least four to six weeks of BAHA CROS use. Tests comprised sound localization and speech reception in noise.

Sound localization was tested with an array of nine loudspeakers at intervals of 30° azimuth.[2,4] The two outermost loudspeakers were included to avoid edge effects. Low (500 Hz) and high (3000 Hz) frequency narrow-band (1/3 octave) noise stimuli were presented at 65 dB SPL with duration of one second. Correct identification of the active loudspeaker and correct lateralization scores were recorded. The scores obtained in this manner were compared to chance levels of

11% and 50%, respectively During the measurements, the patients were not permitted to turn their head

Table 1 Patient characteristics age, gender, cause of deafness, duration of unilateral inner ear deafness and average pure-tone thresholds ($PTA_{0\,5\,1\,2}$) at the frequencies 500, 1000 and 2000 Hz in the contralateral ear
AC=air conduction, BC=bone conduction, HL= hearing loss, eci= e causa ignota

Patient No	Age (yr)	Gender	Etiology	Duration (yr;mo)	$PTA_{0\,5\,1\,2}$ (dB HL) AC	BC
30	71	F	Sensorineural HL eci	07,05	18	18
31	66	M	Acoustic neuroma	11,06	10	10
32	61	F	Stapedotomy surgery	02,09	27	22
33	58	M	Acoustic neuroma	01,03	10	10
34	37	M	Acoustic neuroma	02,08	2	2
35	63	M	Acoustic neuroma	00,11	13	13
36	29	F	Trauma	25,07	3	2
37	60	M	Acoustic neuroma	14,07	23	23
38	53	F	Acoustic neuroma	09,09	15	8
39	57	M	Noise trauma	25,09	8	8
40	42	F	Acoustic neuroma	07,08	8	8
41	39	M	Acoustic neuroma	02,09	7	7
42	54	F	Sensorineural HL eci	04,10	22	22
43	16	F	Inner ear cholesteatoma	02,02	18	3
44	39	F	Congenital	39,01	7	7
45	29	M	Congenital	29,05	5	3
46	69	M	Acoustic neuroma	00,03	22	22
47	55	F	Sensorineural HL eci	24,05	18	23
48	62	F	Trauma	02,00	13	10
49	51	M	Sensorineural HL eci	29,06	38	15
50	47	M	Trauma	00,08	10	10
51	49	M	Trauma	28,07	12	10
52	42	F	Inner ear cholesteatoma	26,08	13	13
53	58	M	Acoustic neuroma	02,01	7	0
54	51	M	Sensorineural HL eci	02,09	18	18
55	25	F	Sensorineural HL eci	22,01	42	32
56	25	M	Inner ear cholesteatoma	19,02	18	17

Speech reception was measured with short, everyday sentences [12] Spectrally-shaped noise (N) was presented in front of the listener, while speech (S) was presented at + 90°, or separately at - 90° azimuth and *vice versa*. In a subsequent condition, speech and noise were presented in front of the patient. In all the test conditions, the noise level was fixed at 65 dBA and speech reception thresholds (SRTs) were measured with an adaptive tracking procedure [12] We employed the speech-to-noise ratio (S/N ratio) or the noise level minus the SRT Each test

condition was measured twice and the results were averaged. These methods were also used by Hol et al.[6]

Patient questionnaires

Baseline (unaided) and post-intervention BAHA CROS patient outcome data were obtained with Dutch versions of the APHAB, GHABP, IOI-HA and SSD questionnaires. The APHAB and the GHABP were administered in the unaided situation and postoperatively after at least 6 weeks of experience with the BAHA CROS. The IOI-HA and the SSD questionnaires were administered 6 weeks after BAHA CROS fitting.

The APHAB consists of 24 items assigned to 4 domains: ease of communication (EC), listening under reverberant conditions (RV), listening in background noise (BN) and aversiveness of sound (AV). Differences between baseline and post-intervention outcomes were used to evaluate patient satisfaction.[8]

The GHABP evaluates initial hearing disability, handicap, hearing aid use and benefit, residual disability and patient satisfaction with hearing aids.[9] Average scores are calculated for each domain and values are scaled to lie between 0 (no problem) and 100 (greatest disability/handicap). Some of the domains (use, benefit and satisfaction) are scored the other way around (0= poorest outcome, 100= best outcome).

The IOI-HA can be applied to make clinical evaluations of hearing aid fitting using a 5-point scale.[13] It consists of seven items about use, benefit, residual activity limitations, satisfaction, impact on others, and quality of life. The highest score (i.e. 5) represents the best outcome. We added two questions: a) Would you recommend the BAHA to someone else with the same type of hearing loss? b) Would you still opt for a BAHA CROS if you had to finance the device by yourself?

The SSD questionnaire consists of 12 questions on use, satisfaction, aesthetics, handling of the BAHA and estimation of hearing aid benefit in different listening situations in comparison with the situation without a hearing aid. Answers can be given on 4-point and 3-point scales. Patients were also asked to give their opinion about the most favourable listening situation (unaided , BAHA or no preference) in 5 specific conditions.

Analysis

Student's t test was applied to the speech reception and localization scores and to the mean values in the different domains of the APHAB and GHABP. A p value of < 0.05 was chosen as the level of significance, with $p < 0.025$ in the case of a two-

tailed t-test Mann-Whitney test was applied as a nonparametric test when n <30 The results were computed using the SPSS package (version 12)

Results

Audiometric measurements

Sound localization

Table 2 shows the sound localization results in response to 500 and 3000 Hz noise stimuli Although the percentages of correct answers in the unaided and BAHA CROS situations were close to chance level they were significantly better than that level of 11% ($p < 0.05$) Mean lateralization scores were not statistically significantly different from the chance level of 50% Scores obtained in the unaided situation were not significantly different from those obtained after BAHA CROS fitting (i e there was no improvement with the BAHA)

The 5 patients with congenital deafness had remarkably higher scores in all the test conditions than the patients with acquired deafness Significant differences in response to the 500 and 3000 Hz stimuli were computed in the unaided situation between the congenital and acquired group ($p< 0.05$) No statistically significant differences in improvement were seen after BAHA fitting (aided minus unaided) between the various subgroups ("acoustic neuroma" n=28, "congenital" n=5, "other" n=23)

Table 2 Average sound localization and lateralization scores (i e correct identification of the active loudspeaker) in response to 500- and 3000-Hz noise band stimuli * Significantly better than chance level

Identification	Chance level	Frequency Hz	Unaided (n=27)	BAHA (n=27)	Unaided (total n=56)	BAHA (total n=56)	BAHA (Hol et al n=29)
Correct (%)	11	500	20 7*	20 8*	19 8*	18 5*	16 5*
		3000	20 6*	18 2*	19 1*	17 8*	17 4*
Lateralization (%)	50	500	55 8	52 2	50 1	49 1	46 2
		3000	54 8	53 3	52 0	53 1	52 6

Figure 1A

Figure 1B

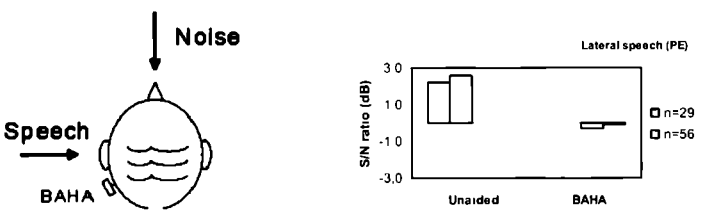

Figure 1 A/B: S/N ratios that yielded speech intelligibility scores of 50% in response to everyday Dutch sentences when the noise was presented in front of the patient and the speech on the side of the poor ear (PE) or best ear (BE): `lateral speech` unaided and with the BAHA CROS (BAHA) * $p < 0.025$, compared to the unaided situation

Speech reception

Lateral speech and frontal noise

Figure 1 shows the mean S/N ratios in the unaided and the BAHA CROS situations when the noise was presented in front of the listener and the speech on the side of the poor ear (PE) or best ear (BE). Better performance corresponds with a lower S/N ratio. In the unaided situation the difference in S/N ratio when the speech was presented to PE or BE (i.e. the head shadow effect) was 5.2 dB. This effect decreased to 3.0 dB with the BAHA CROS. Therefore, the BAHA CROS reduced the head shadow effect by 2.2 dB, which was equivalent to an increase of 33% in speech recognition in this particular test condition.

Lateral noise and frontal speech

Figure 2 shows the results when the speech was presented in front of the listener and the noise to the PE or BE. In the unaided situation, the S/N ratio was –2.8 dB when the noise was presented to the PE.

Figure 2A

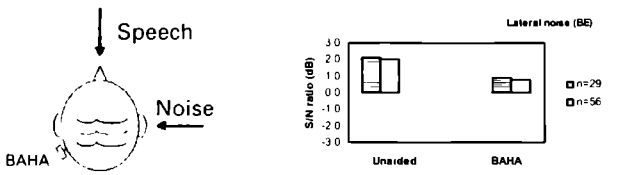

Figure 2B

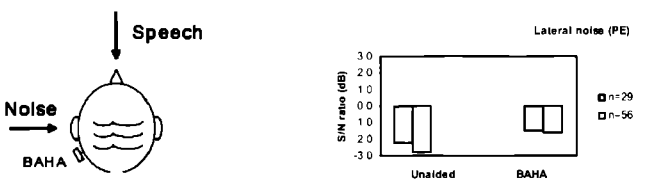

Figure 2 A/B: S/N ratios that yielded speech intelligibility scores of 50% in response to everyday Dutch sentences when the speech was presented in front of the patient and the noise on the side of the poor ear (PE) or best ear (BE): `lateral noise` unaided and with the BAHA CROS (BAHA) * $p < 0.025$, compared to the unaided situation.

With the BAHA CROS, the S/N ratio increased (deteriorated) with 1.2 dB. When the noise was presented to the BE in the BAHA CROS situation, the S/N ratio decreased (improved) with 1.2 dB (18% improvement in speech recognition).

Frontal speech and frontal noise

When the speech and noise were presented in front of the patient in the unaided and in the BAHA CROS situation, the S/N ratios were essentially the same (–0.2 dB). Significant changes are indicated in table 3.

In 4 conditions, the performance of the congenital patients was significantly better than that of the acquired group: lateral noise with the BAHA on the poor side ($p=0.04$), lateral speech unaided in the best ear ($p=0.04$), lateral speech with the BAHA in the best ear ($p=0.01$) and frontal speech and noise unaided ($p=0.01$). No significant differences were seen between the acquired deafness subgroups ("acoustic neuroma n=28, "others" n=23).

Table 3. S/N ratios of the total group (n=56) in the unaided and BAHA CROS situations in response to different speech and noise configurations. NH refers to the side of the head with the normal ear and Deaf refers to the impaired side.

Lateral Noise				Lateral Speech				Frontal speech/Noise	
Unaided		BAHA		Unaided		BAHA		Unaided	BAHA
NH	Deaf	NH	Deaf	NH	Deaf	NH	Deaf		
2.0	-2.8	0.8*	-1.6**	-2.6	2.6	-3.1	-0.1*	-0.2	-0.2

*Significant improvement compared to the unaided situation ($p < 0.025$)
**Significant deterioration compared to the unaided situation ($p < 0.025$)

Patient questionnaires

APHAB

A total of 47 patients completed the AHPAB questionnaire (84%). APHAB scores are shown in Figure 3. Lower scores denote improved communication. BAHA CROS fitting led to significant improvements ($p< 0.025$) in 3 domains: ease of communication, background noise and reverberation. Most improvement was seen in the Background Noise (BN) domain.

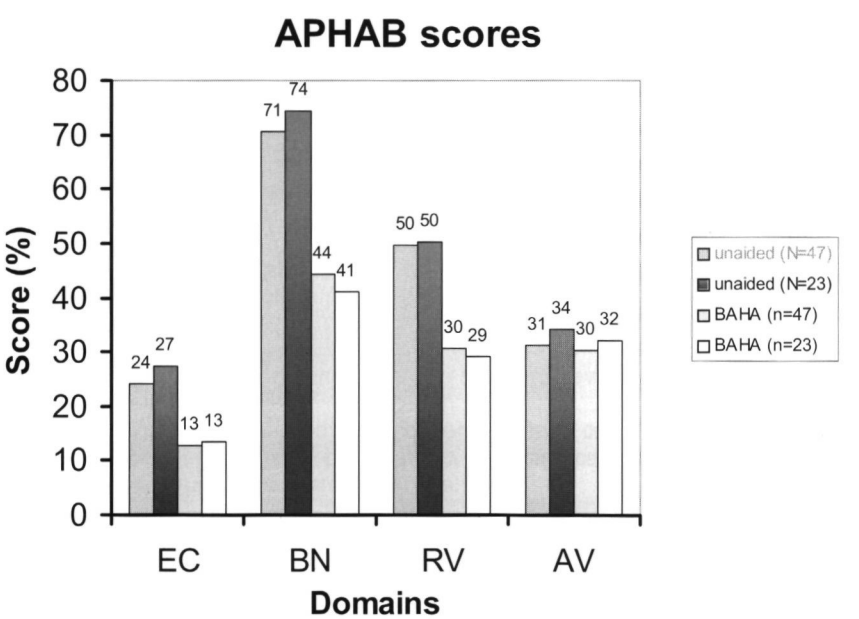

Figure 3. Mean APHAB scores of the previous 23 patients[6] and a total of 47 patients in the domains Ease of Communication (EC), Background Noise (BN), ReVerberation (RV) and AVersiveness of sound (AV) in the two different situations: unaided and with the BAHA CROS. Lower scores reflect improved communication.

Scores in the domain AVersiveness of sound (AV) were around the same with the BAHA CROS These data were comparable with those reported in the previous study of Hol et al The mean scores of the 5 patients with congenital unilateral inner ear deafness in the 4 domains were not significantly different from those of the acquired group Nevertheless it was interesting to note that in the unaided and BAHA CROS situations, the congenital patients had more favourable scores on the domain ease of communication

GHABP

The response rate to the pre- and post-intervention questionnaires was 84% (n=47) Figure 4 shows the GHABP unaided situation scores Mean initial disability and unaided handicap scores were 53% and 39%, respectively Figure 5 shows utility, benefit, residual disability and satisfaction with the BAHA CROS in the total group (n=47) and in the original series of patients (n=23) In the total group mean day-to-day use of the BAHA CROS was 84% Mean benefit and satisfaction were 51% and 49%, respectively Residual disability was 37% in the total group No statistically significant differences in scores were found between the congenital group and the acquired group

IOI-HA

The response rate to the IOI-HA questionnaire was 88% (n=49) The seven items are use (1), benefit (2), residual activity limitations (3), satisfaction (4), residual participation restrictions (5), impact on others (6), and quality of life (7)

Responses are scored on a five-point scale and a higher score represents a better outcome Mean scores on the seven items were 4 4, 3 4, 3 7, 3 9, 4 0, 4,3 and 3 9, respectively (Table 4) Our additional items revealed that 90% (n= 43) would recommend the BAHA to someone else with the same type of hearing loss and 69% (n=34) would still opt for a BAHA CROS if they had to finance the device themselves Only one patient (2%) gave a negative answer to this second additional question

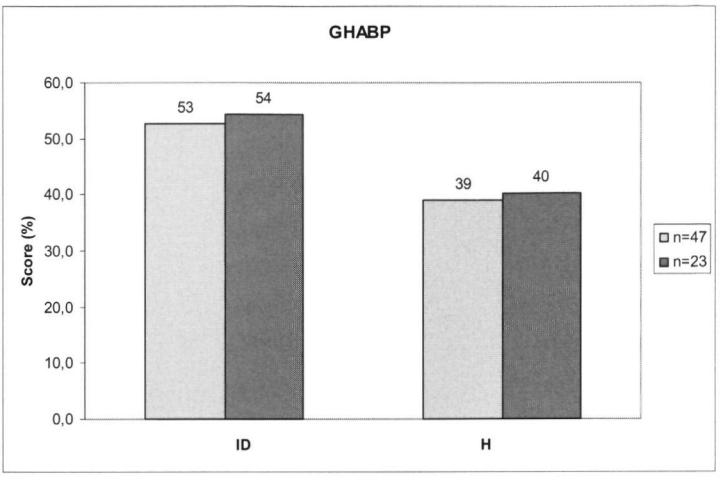

Figure 4. Mean GHABP scores in the unaided situation in the domains Initial disability (ID) and Handicap (H) of the previous 23 patients[6] and the total group (n=47) (0= best outcome, 100 poorest outcome)

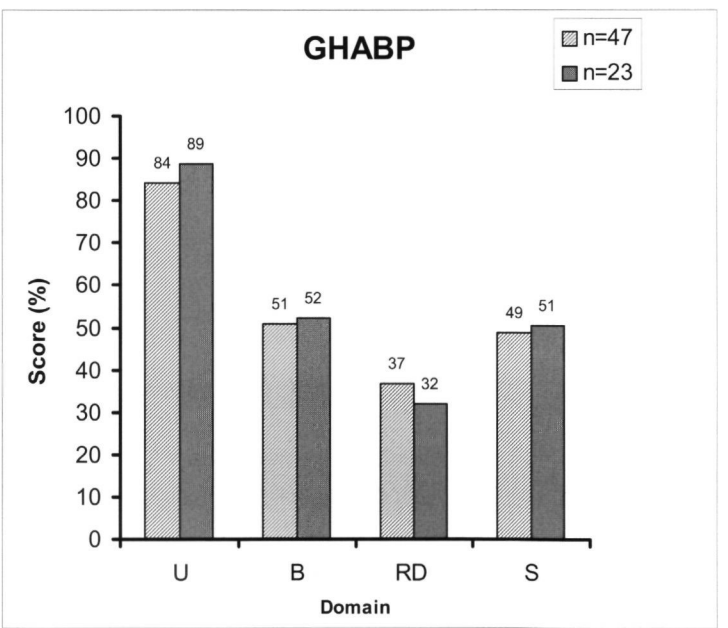

Figure 5. Mean GHABP scores of the previous 23 patients[6] and the total group (n=47) in the domains Utility (U), Benefit (B), Residual Disability (RD) and Satisfaction (S) with the BAHA CROS (0= poorest outcome, 100= best outcome)

Table 4 Mean IOI-HA scores with the BAHA CROS. Higher scores reflect better outcomes Responses were given on a 5-point scale. Items: use (1), benefit (2), residual activity (3), limitations (4), satisfaction (5), impact on others (6) and quality of life (7)

IOI-HA							
Item	1	2	3	4	5	6	7
Hol et al (n=23)	4.6	3.5	3.9	4.0	4.2	4.5	4.1
Total (n=49)	4.4	3.4	3.7	3.9	4.0	4.3	3.9

SSD questionnaire

A total of 51 patients responded to the SSD questionnaire (91%). Thirty-eight patients used the BAHA every day for more than 8 hours a day (75%). Only one patient was using the BAHA for less than 2 hours a day. In 36 patients (73%) quality of life had improved since BAHA fitting. Satisfaction with the BAHA was on average 7.4, scored on a 10-point scale. The mean score on BAHA aesthetics was 7.7. In 5 specific listening conditions, the patients were asked to give their preference, for unaided, BAHA, or no difference. In quiet, 32 patients (64%) stated that the BAHA was better than the unaided situation, while 17 patients (30 %) had no preference. When speaking to a person in a group, listening to music, listening to television and when sitting at the dining table with someone speaking on the impaired side, the BAHA was preferred by 72% (n= 36), 70% (n=35), 84% (n=42) and 84% (n=42), respectively. In the subjective ability to localize sounds, there was no clear preference for the BAHA: 12 patients (24%) preferred the BAHA, 11 patients preferred unaided, 18 patients indicated the unaided and aided situations and 10 patients stated 'no difference'.

When we took a closer look at patients nos 49 and 55 who had subnormal hearing in the contralateral ear, we found that their audiometric and subjective results did not differ significantly from those of the total group ($p>0.05$).

A total of 5 patients did not return the SSD questionnaire despite several reminders. Further enquiries revealed that one of these 5 non-responders had stopped using the BAHA, because it had provided insufficient benefit.

Discussion

Unilateral inner ear deafness is a serious handicap that has strongly deleterious effects on communication in difficult listening conditions. Especially patients with acquired deafness are well-aware of the differences compared to their previous experience of binaural hearing. It is a challenge to find adequate solutions to

minimize this handicap. Outcomes with the conventional CROS hearing aid are generally rather disappointing, so it is worthwhile to evaluate new options. One of these options is the BAHA CROS. Clinical evidence about the value of the BAHA CROS treatment will increase on the basis of step-by-step clinical results from an increasing number of consecutive patients (n=56) with unilateral inner ear deafness. Comparisons can be made with the unaided situation, including evaluations of subjective benefit. Previous studies on speech reception in noise and subjective auditory abilities showed that the BAHA CROS was more advantageous than the conventional CROS and the unaided situation.[7] Nevertheless a well-structured audiological test setting with the head kept in a fixed position did not show any significant improvements in auditory localization.

Results of the 56 BAHA-CROS patients in this extended series were in agreement with the results previously published by Hol et al. Therefore, BAHA CROS application is having consistent positive effects in the Nijmegen series.[6] This success may have been influenced by the etiologies of deafness, the duration of unilateral total inner ear deafness and selection procedures. Hol et al reported that 9 out of 39 patients declined BAHA CROS application after a preoperative trial period with the BAHA on a headband. About 25% of the patients experienced insufficient benefit and withdrew from the implantation programme. It would be interesting to evaluate the personal and social characteristics of these patients to obtain information about the profile of patients who benefit from BAHA application. In a recent study by Andersen et al,[14] 59 patients with unilateral hearing loss after acoustic neuroma surgery were evaluated for BAHA treatment. Half of the patients accepted a trial period with the BAHA on a headband, but only 25% ultimately opted for implantation.

In our study group no exact data are available on the patients, who declined implantation after a trial period with the BAHA on a steel headband. It is our intention to gain more insight into the arguments of these patients. Further research into this issue is in progress.

The benefit of reducing the head shadow effect was again confirmed with speech recognition measurements and showed an increase of 33% in speech recognition in the situation with speech coming from the poor ear.

The initial disability and hearing handicap scores obtained from our patients using the GHABP (53% and 39%, respectively) were considerably lower than the scores obtained from BAHA patients with the conventional indication for BAHA treatment (75% and 82%, respectively).[15] In the study by Andersen et al,[14] 45% of the patients reported that they had a significant hearing handicap, while 38 %

reported a moderate level of handicap. There was no significant correlation between a significant high handicap and positive interest in implantation.
In our series it was remarkable that only 5 out of a total of 56 patients had congenital deafness. Their sound localization scores in the unaided situation were significantly better than those of the acquired deafness group. Also their speech reception scores were significantly better than the acquired group. This might be partially explained by the relatively good air conduction thresholds in their contralateral ear (mean 9.2 dB HL).
There was however no significant difference in subjective benefit between these two groups. The subjective handicap and initial disability scores measured by the GHABP were assigned equal scores. In the 5 congenital patients, these scores were 57% and 41%, respectively. Apparently, the congenital patients were able to make certain adaptations to achieve directional hearing. Nevertheless they reported the same levels of disability and handicap.
BAHA application did not lead to significantly better results in the subgroup of acoustic neuroma patients. However, patient satisfaction seemed to be higher in this patient group and they had significantly better scores in the GHABP domains benefit, satisfaction with the BAHA CROS and residual handicap.
In the literature, many BAHA CROS patients have an etiology of acoustic neuroma. The Nijmegen series also contains a high number of acoustic neuroma patients: 17 among the first 29 patients (Hol et al)[6] and 10 in the subsequent 28 patients. When we started to compile this series, we first selected the acoustic neuroma patients from our clinical files for a trial period with the BAHA on a steel headband. This precautionary approach was intended to create a patient series in which BAHA CROS application would have a high degree of success, in order to provide clinical evidence of the value of this treatment. In the meantime, BAHA CROS application has been approved in the Netherlands and by the Federal Drug Administration in the USA, based on the positive outcomes at European and American BAHA centres.
By extending the series, we found additional proof that the treatment is effective. Originally, we used strict patient selection criteria: profound inner ear deafness on one side and (almost) normal hearing (thresholds at or better than 25 dB HL) on the contralateral side with a negligible air-bone gap (<10 dB HL). In this extended series, two patients did not meet these criteria exactly (patient no. 49 with an air-bone gap of 23 dB and patient no. 55 with air and bone conduction thresholds of 42 dB and 32 dB, respectively). No significant differences in audiometric and subjective outcomes were found between these patients and the rest. Comparable results were found by Vaneecloo et al who used less strict criteria, such as mixed

hearing loss on the better side and some profitable hearing on the impaired side.[5] Therefore, it may be possible to broaden our inclusion criteria somewhat.

In conclusion, consistent positive effects of BAHA CROS application were observed in our extended series of 56 BAHA CROS patients (we excluded 2 non-users). To obtain more information about patient profiles that may benefit from BAHA CROS application, more research is needed.

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4.2

Randomized controlled trial on the effectiveness of the conventional CROS, the transcranial CROS and the BAHA transcranial CROS; a randomized controlled trial in adults with unilateral inner ear deafness

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Abstract

Objective: To evaluate the effectiveness of three CROS hearing aids in adults with unilateral inner ear deafness.
Study design: Prospective randomized controlled trial.
Settings: Tertiary referral centre.

Subjects: Ten patients with unilateral inner ear deafness and normal hearing in the contralateral ear were selected to evaluate three different methods of amplification: the conventional contralateral routing of sound (CROS) hearing aid, the completely-in-the-canal hearing aid (CIC) and the bone-anchored hearing aid CROS (BAHA). Two patients had congenital unilateral hearing loss, the other eight patients had acquired unilateral hearing loss due to acoustic neuroma excision (n=1), trauma (n=3), meningitis (n=2) and sudden deafness with cause unknown (n=2).

Methods: The patients tried each of the three hearing aids in a random order for a period of eight weeks. Audiometric performance, including speech in noise, directional hearing and subjective benefit were measured after each trial period, using the APHAB, SSQ and single sided deafness questionnaire. Unaided baseline measurements were used for comparison purposes to assess the effectiveness of each of the three amplification methods.

Results: Sound localization performance was essentially at chance level in all four conditions. The APHAB showed subjective improvement, particularly seen with the BAHA CROS. Mixed results were seen on the other patient outcome measures that alternated in favour of one of the three CROS devices. After the trial, six patients out of the ten patients decided not to apply for any of these CROS hearing aids, three chose to be fitted with the BAHA CROS and one with the conventional CROS.

Conclusions: Most of the patients experienced some degree of benefit with each of the three hearing aids, but it was not large enough to outweigh the dis-advantages. Preference for one of the three hearing aids was independent of the order in which they were tried. We recommend that all patients with unilateral inner ear deafness who apply for hearing revalidation should be offered a trial with at least the BAHA CROS. It would be worthwhile to formulate selection criteria to help establish which hearing aid will provide the most benefit.

Introduction

Before considering to fit a hearing aid in patients with single sided deafness (SSD), it is important to investigate their medical and social backgrounds Harford and Dodds[1] were among the first who advocated contralateral routing of sound (CROS) amplification for such patients, in order to eliminate head shadow These authors found that the degree of success depended on the motivation of the patient and the listening demands imposed by their lifestyle and working environment They applied what is nowadays known as the "conventional CROS" device It comprises a microphone placed near the impaired ear and an amplifier (hearing aid) near the normal ear The signal is presented to the normal ear via an open ear mould Conventional CROS devices only transmit the frequencies of above 1000 Hz through this open ear mould, which results in a "tinny" sound that might help the patient to localize sounds Acceptance of this conventional CROS device by the patients was related to the level of hearing in their best ear When hearing was within normal limits, the success rate was low only one out of 12 patients accepted the CROS device However, when mild high frequency hearing loss was present, the success rate was increased to 54% [1]

Lotterman and Kasten[2] studied the effect of a conventional CROS device when words were presented against a background of cafetena noise They observed favourable results when the speech was presented near the impaired ear, but unfavourable results when the speech was presented near the normal ear Markides[3] reported similar results He also tested directional hearing, but found that none of his patients could localize sounds

With the conventional CROS, sound is received on the impaired hearing side and transmitted by a cord around the neck, or by wireless FM transmission, to the best ear [4 5] However, many patients find it unpleasant to have an ear mould in their best ear and a cord around their neck The ear mould causes at least partial occlusion of the best ear, which is a key consideration, because the adaptive behaviour learned by individuals with unilateral hearing loss often involves turning the best ear towards the sound source

Another option, is the use of bone conduction in the form of a "transcranial CROS" device In 1960, Fowler[6] suggested the use of a bone conduction hearing aid (in a spectacle frame) near the deaf ear This would stimulate the normal cochlea (cross stimulation) by bone conduction i e through the skull No results of this application were presented at that time Probably the first, comprehensive, study on transcranial CROS via bone conduction was published in 1991 by Welling and co-workers [7] They used the implantable Audiant Bone Conductor and reported

positive results in some of their patients. One of the main limitations of the device was insufficient output.[8]

In 2000, Vaneeclo et al.[9] applied the more powerful BAHA (bone-anchored hearing aid) as a transcranial CROS device in patients with unilateral inner ear deafness. High patient satisfaction was reported as well as improvements in directional hearing. Other studies on motivated patients also showed high patient satisfaction, but the improvements in directional hearing could not be replicated, which drew attention to the sound localization problems in patients with unilateral inner ear deafness.[10-12] Good speech-in-noise results reflected the benefit of a BAHA CROS in lifting the head shadow while the compact design avoided some of the disadvantages of a conventional CROS. Therefore, the BAHA CROS is becoming more and more popular in patients with unilateral inner ear deafness.[13]

Another option for patients with unilateral inner ear deafness was introduced in the late nineteeneighties.[14] It comprised a high power conventional air-conduction hearing aid with a relatively long ear mould that fit deeply into the patient's impaired ear and left the best ear unoccluded. When the amplified signals were loud enough they caused vibration of the bony walls of the ear canal and middle ear, which stimulated the normal ear by means of bone conduction through the. To achieve substantial gain and to overcome the interaural attenuation of the skull, very tight fitting is required deep within the bony ear canal. This application therefore forms an alternative transcranial CROS device that is fitted completely-in-the-canal (CIC). Valente et al.[15] reported a success rate of 50% in their patients, which was high in comparison with their success rate of 10% with conventional CROS application. The improvement was ascribed to better sound quality: the harshness or tinny sound was gone. Hayes and Chen[16] also reported on the CIC device, but their study group only comprised three cases. As far as we know, very little attention has been paid to this type of transcranial CROS method since then.[14-17]

Faced with the increasing demands of communication skills in modern society, professionals are being urged more and more to recognize the detrimental effects of unilateral inner ear deafness. Currently these patients can choose to learn various coping strategies, or they can give preference to receiving unilateral amplification by means of a CROS hearing aid. Little has been published about the conventional CROS hearing aid or the completely in the canal (CIC) device. Although mostly poor results have been reported in the few available studies, some patients do benefit from CROS application.

Recently, many patients with unilateral sensorineural hearing loss become aware of the BAHA CROS option, not least due to its the high profile on the internet.[18]

Baguley et al reviewed the published studies in order to emphasize the need for evidence base for the application of the BAHA CROS in these patients with single sided deafness [18] The reviewed studies found evidence of improved performance with the BAHA CROS in patients with unilateral profound hearing loss and (near-) normal hearing in the contralateral ear [10 11 19] However, Baguley et al criticized these studies on methodological aspects [18] All the studies, however, recommended careful selection of patients for the BAHA CROS as it was particularly advantageous in specific listening situations, but did not lead to objectively measurable improvements in directional hearing Baguley et al therefore advised clinicians to proceed with caution and to await the outcome of a larger randomized trial [18] The largest series of patients with unilateral sensorineural hearing loss who have been fitted with a BAHA CROS recently reached 56 [20] These patients were selected on the basis of a pre-operative trial with the BAHA CROS on a headband On average, the majority of these 56 patients are satisfied [20]

On account of the criticism from Baguley, we performed a pilot study in which the three currently available CROS devices (Conventional CROS, completely-in-the-canal -CIC- and BAHA CROS) were assigned in random order to ten adult patients with unilateral sensorineural hearing loss for a test period of 8 weeks

The evaluation comprised audiometric testing and patient outcome measurements on three hearing specific instruments to quantify subjective benefit

Patients and methods

Patients

At our outpatient clinic, we recruited ten adult patients with unilateral sensorineural hearing loss These patients had not necessarily visited our outpatient clinic to obtain information about hearing aids They were invited to participate in a prospective trial with three different methods of unilateral amplification the conventional CROS (CROS), the completely-in-the-canal (CIC) and the BAHA CROS on a headband (BAHA)

Each subject's unaided performance provided a baseline measurement to assess the effectiveness of the CROS devices All the patients had normal hearing (PTA< 25 dB) in the contralateral ear with a mean PTA of 12 dBHL. Two patients had congenital unilateral hearing loss, the other eight patients had acquired unilateral hearing loss due to acoustic neuroma excision (n=1), trauma (n=3), meningitis (n=2) and sudden deafness with cause unknown (n=2) Average duration of

deafness was 23 years (range 1-56) Table 1 presents an overview of gender, age, aetiology and duration of unilateral hearing loss

Table 1 Patient characteristics gender, age, aetiology, duration of unilateral deafness and average pure tone average (PTA) at the frequencies 500, 1000 and 2000 Hz in the normal ear

Patient	Gender	Age (yrs)	Aetiology	Duration (yrs)	PTA BC	PTA AC
1	M	31	Congenital	31	8 3	13 3
2	M	51	Trauma	41	8 3	8 3
3	M	64	Acoustic neuroma	10	0 0	3 3
4	F	56	Congenital	56	5 0	5 0
5	M	45	Trauma	1	8 3	8 3
6	F	28	Meningitis	27	13 3	18 3
7	M	47	Trauma	3	23 3	25 0
8	F	44	Sudden deafness	1	13 3	13 3
9	F	53	Meningitis	41	13 3	13 3
10	M	43	Sudden deafness	18	8 0	8 0

AC, air conduction, BC bone conduction

Methods

Patients were offered a trial period of 8 weeks with each of the three CROS devices for a period of 8 weeks in a random order The conventional CROS consisted of a behind-the-ear hearing aid with a wire around the neck (Widex B2 with Widex CROS unit on the contralateral side) The Beltone P60PP completely-in-the canal (CIC) hearing aid, deeply fitted, was tested after a patient-specific mould had been made for the ear canal Fitting parameters of both devices were set according to the specifications of the manufacturer The BAHA CROS was worn on a steel headband Patients were encouraged to use the devices on a daily basis The CROS devices were fitted without providing any information about efficacy or comfort At the end of the trial, the patients were asked whether they felt that the unilateral amplification had been worthwhile and which of the three CROS devices (if any) took their preference

Evaluations were made in four different conditions unaided, with the conventional CROS, with the CIC and with the BAHA CROS on a headband

The data obtained in the unaided condition served as baseline measurement to assess the effectiveness of the hearing aids Afterwards, the patients were fitted with 1 of the 3 devices for eight weeks In all four conditions, the audiometric evaluation consisted of sound localization measurements with a 9-speaker array at 30° [for details see Bosman et al[21]] Speech perception was measured using

short, everyday Dutch sentences [22] Spectrally shaped noise at a fixed level of 65 dB was presented in front of the listener and speech at + 90°, - 90° azimuth and vice versa Speech reception thresholds were measured with the "one up-one down" adaptive tracking procedure described by Plomp and Mimpen [22]

Patient outcome measures

The baseline (unaided) and post-intervention measurements were conducted using 2 validated instruments in the Dutch language the Abbreviated Profile of Hearing Aid Benefit (APHAB)[23] and the Speech, Spatial and Qualities of hearing profile (SSQ) [24] Patients were asked to fill out the post-intervention instruments after 8 weeks of experience with each device

The Abbreviated Profile of Hearing Aid Benefit (APHAB)[23] consists of 24 items in the following domains ease of conversation (EC), listening under reverberant conditions (RV), listening in background noise (BN) and aversiveness to loud sounds (AV) Higher scores reflect more problems

The Speech Spatial and Qualities of Hearing scale (SSQ)[24] measures benefit on the domains spatial hearing and speech perception As the spatial hearing domain is presumed to be of importance to binaural hearing, we used the 16 items from the spatial hearing domain of the questionnaire (e g locate speaker round a table, locate dog barking, judge distance of a vehicle, etc) Patients rated each item on a 10-point Likert scale Higher scores reflected good performance

The 12-item single sided deafness (SSD) questionnaire[25] was also administered to obtain data on use, satisfaction, estimation of hearing aid benefit in different listening situations in comparison with the unaided situation, aesthetics and handling of the CROS devices This questionnaire has been used in previous studies on unilateral inner ear deafness [25]

Results

General

One patient did not participate in all unaided audiometric measurements Two patients could not be fitted with the CIC, due to an enlarged external meatus secondary to tumour excision One of these two patients dropped-out, so CIC and CROS measurements were lacking Another patient tried all three hearing aids, but was unable to complete the audiometric testing In addition for this patient most of the questionnaires were too difficult to fill out adequately, even with

assistance. All the patients filled out the patient outcome measures in nearly all the conditions. When data were missing, this is specified.

Source localization

Sound localization performance was essentially at chance level in all four conditions. Data on lateralization (left/right) scores are shown in Table 2.

Table 2. Average sound lateralization scores using 500 and 3000 Hz stimuli in four conditions: unaided, conventional CROS (CROS), completely-in-the-canal (CIC) and BAHA CROS (BAHA). Chance level for lateralization (50%) is shown

	Chance level %	Frequency Hz	Unaided %	CROS %	CIC %	BAHA %
Lateralization	50	500	53.6	53.0	53.3	56.1
		3000	61.0	48.6	70.4	58.9

Speech recognition

Speech reception thresholds (SRTs) in noise in all four conditions are shown in Figure 1. Better performance corresponds with lower signal-to-noise ratios. Data were collected with the noise in front, while the speech was presented on either the profoundly deaf side (PE) or the normal hearing side (BE). This condition was called lateral speech. In the unaided condition, (see figure 1) the S/N ratio when the speech was presented on the deaf side with the noise in front, was about 0.5 dB. In the aided conditions, these S/N ratios were better with the conventional CROS (reduced to about -1.7 dB), slightly poorer with the BAHA CROS (0.4 dB) and somewhat poorer with the CIC (about 0.7 dB).

When the speech was presented on the normal side (best ear = BE), the S/N ratio in the unaided condition was -4.3 dB. With the conventional CROS, the CIC and the BAHA CROS, these S/N ratios were -2.7 dB, -4.6 dB and -2.0 dB, respectively.

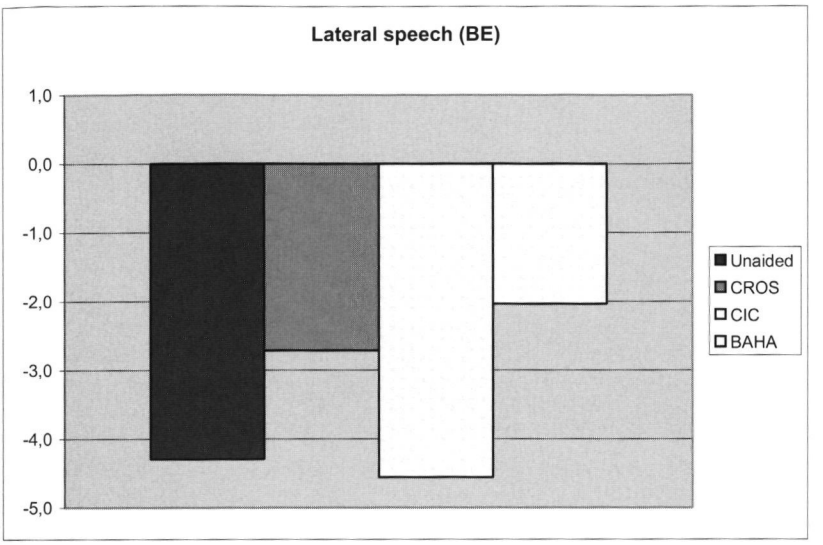

Figure 1A

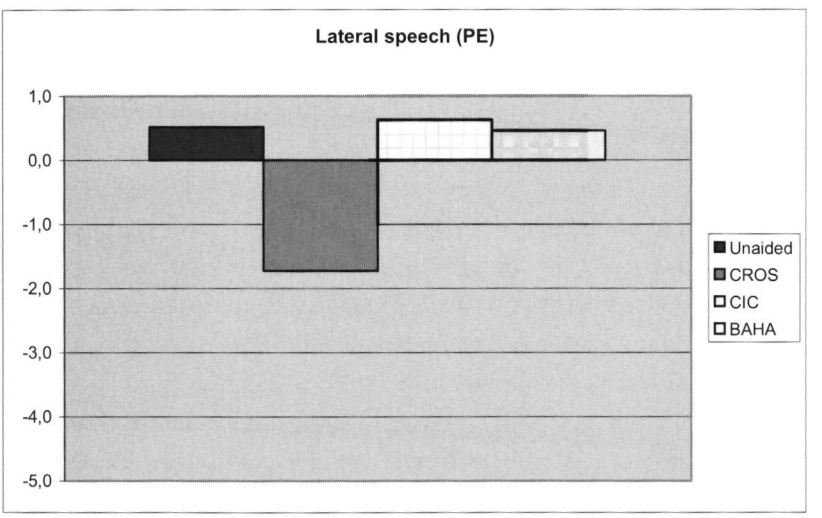

Figure 1B

Figure 1.
A/B S/N for a speech intelligibility of 50% with everyday Dutch sentences: with noise presented in front and speech on either the poor ear (PE) or the best ear (BE). 'Lateral speech' in four conditions: unaided, conventional CROS (CROS), completely in the canal (CIC) and BAHA CROS (BAHA)

Outcome measures

APHAB

Scores on the different domains of the APHAB are shown in Figure 2.

The conventional CROS showed improved scores on the domains EC (22.1), BN (60.6) and RV (38.4). All three hearing aids showed deterioration on the domain AV. The least deterioration was seen with the conventional CROS (39.6).

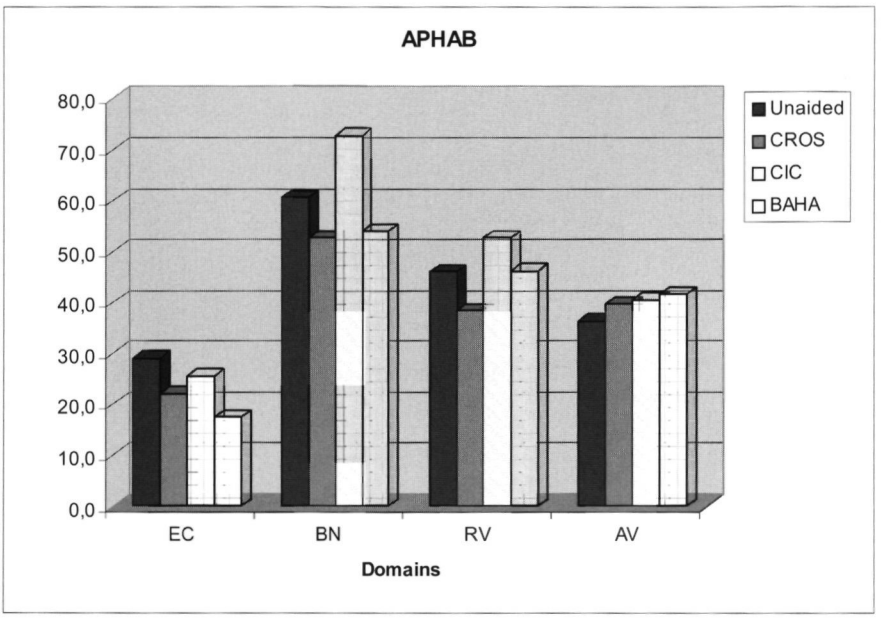

Figure 2. Mean scores of the 10 patients on the APHAB in the domains Ease of Communication (EC), Background Noise (BN), ReVerberation (RV) and Aversiveness of sound (AV) of the APHAB in four different conditions: unaided, conventional CROS (CROS), completely-in-the-canal (CIC) and BAHA CROS (BAHA).

The CIC showed an improved score on the domain EC (25.6), but poorer scores on the other domains BN (72.9), RV (52.7) and AV (39.6).

The BAHA CROS showed the largest improvement on the domain EC (17.7) and less pronounced improvement on the domain BN (54.0). The scores were poorer on the domains RV (46.2) and AV (41.7). Overall, the conventional CROS had the best scores on the APHAB domains.

SSQ

Results are presented in Table 3. The mean score on the spatial domain in the unaided condition was 3.7 on a scale from 0 to 10. In the aided conditions with the CROS, CIC and BAHA the mean scores were better: 1.3, 0.3 and 1.1, respectively.

Table 3: Mean scores of the ten patients on the spatial domain of the SSQ in the four conditions. unaided, conventional CROS (CROS), completely-in-the-canal (CIC) and BAHA CROS (BAHA)

SSQ	Mean	Difference
Unaided	3.7 (1.5)	.
CROS	5.0 (1.8)	+ 1.3
CIC	4.0 (1.4)	+ 0.3
BAHA	4.8 (2.5)	+ 1.1

SSD

Most of the patients used each of the CROS devices more than 8 hours a day (CIC), or 4 to 8 hours a day (CROS, BAHA), 6 to 7 days a week. The CIC showed the highest subjective opinion on a scale from 0 to 10 on the domains wearing comfort, easy to use, rustle, whistle and failure. Lower scores were assigned to the conventional CROS and the BAHA CROS. However, most of the patients (n=6) said that the BAHA CROS was beneficial to hearing, whereas the CIC was of no benefit (n=6) (see also figure 3). The average score on several quality of sound items was best with the CROS (7 on a scale from 0 to 10), slightly lower with the BAHA (6.8) and very poor with the CIC (3.7).

After completion of the trial, the patients were asked whether they wished to apply for one of the three CROS devices. Six patients declined, three patients opted for a BAHA CROS (nos 6, 8 and 10, see Table 1) and one patient chose the conventional CROS with FM link (no. 3, see Table 1). The first two BAHA CROS patients have been implanted and they are satisfied with its performance. The third patient is awaiting surgery. None of the patients who participated in our trial chose the CIC.

Discussion

In this trial three different unilateral amplification options were tested by patients with unilateral inner ear deafness. Their experience was intended to act as an evidence base for patients with difficulties in daily life who are looking for a solution. The BAHA CROS has been found to alleviate the head shadow effect.[10,11 19,26] The conventional CROS is known to have its own merits, but also disadvantages.[27] The completely-in-the-canal hearing aid (CIC), an alternative transcranial CROS application, has received only sparse attention in the literature.

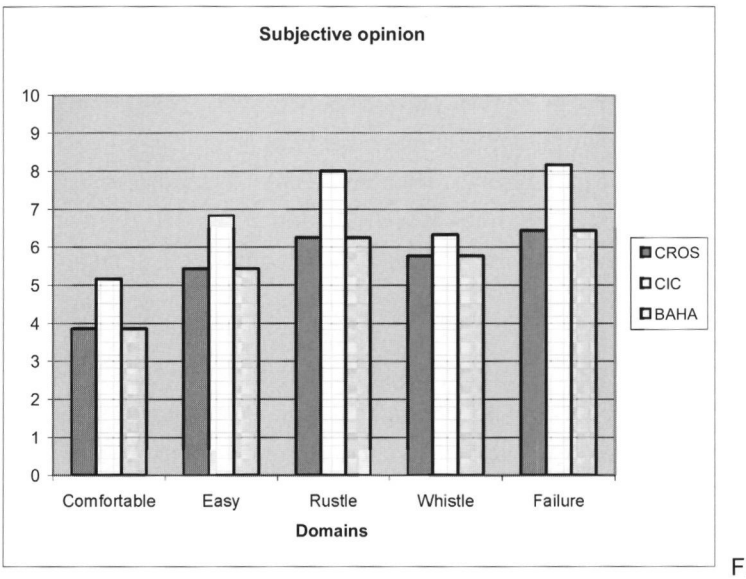

Figure 3A

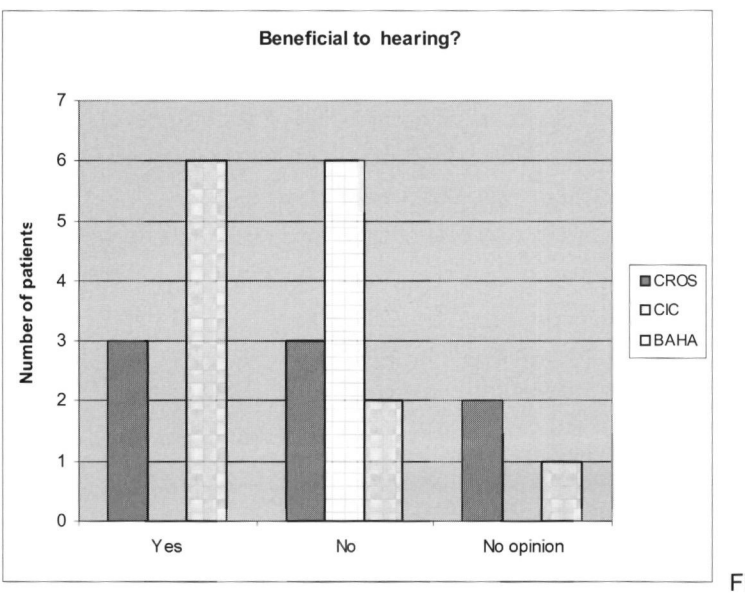

Figure 3B

Figure 3

A Single sided deafness (SSD) questionnaire results. Subjective benefit with each hearing aid was scored in five domains (wearing comfort, easy to use, rustle, whistle and failure) on a scale from 0 to 10

B Number of patients who indicated benefit with one of the hearing aids.

The BAHA CROS is the only system that requires osseo-integration and therefore surgery in preparation for fitting. However, it can also be tested pre-operatively by

means of a transcutaneous set-up in which the BAHA is connected to a special plastic disc held in place by a steel spring headband If the patients are satisfied with this transcutaneous application, they will experience even more benefit from the ultimate bone conduction system after implant surgery

This pilot study investigated the patients' experience with three different CROS devices that they tried in a random order for equal periods of time An evidence base is required for these treatments in patients with unilateral sensorineural hearing loss [28 29]

The directional hearing measurements obtained in this study confirmed our previous findings in patients with unilateral deafness the results were essentially around chance level, irrespective of which device they were using [10 25] In our set-up, the patients were instructed to keep their head facing the front Interestingly, the scores at 3000 Hz were somewhat poorer with the CROS and BAHA than in the unaided condition, but there was a small improvement with the CIC However, none of the scores were statistically significant, probably due to our small numbers Nevertheless, this was the first random controlled trial that not only evaluated the conventional CROS and the BAHA CROS, but also the CIC In our opinion, the direction of our results is of great value Furthermore, the sound localization results have been confirmed in studies with larger numbers, binaural hearing cannot be achieved according to our measurements [20]

The small numbers were probably also responsible for the diverse results on the SRT It was remarkable that the CIC produced such favourable SRT results, but none of the patients chose the CIC

Patient outcome measures are essential in the evaluation patient benefit We used three different instruments the APHAB, the SSQ and the SSD questionnaire

The APHAB showed the poorest scores with the CIC, the best scores with the BAHA and intermediate scores with the conventional CROS

According to the spatial part of the SSQ, the patients experienced some benefit in directional hearing with the conventional CROS, the CIC and the BAHA The BAHA and the conventional CROS showed the most benefit (overall improvement of 1 1 and 1 3, respectively) In comparison, Noble and Gatehouse obtained a mean unaided score of 4 8 from 50 patients with asymmetrical sensorineural hearing loss [24]

Remarkably, the CIC had the best scores on the SSD items wearing comfort, easy to use, rustle, whistle and failure The average score on several quality of sound

items was best with the conventional CROS (7 0), slightly poorer with the BAHA (6 8) and very poor with the CIC (3 7)
At the end of the trial six out of the ten patients did not choose any of the unilateral amplification methods they tested in this study The patient who chose the conventional CROS is using a FM link instead of a wire around the neck The two implanted BAHA CROS patients are satisfied with the performance of the device, the third patient who chose the BAHA CROS is awaiting surgery
In literature is reported that 25% of the patients who underwent acoustic neuroma surgery apply for implantation of the BAHA system after trial on a headband [30] Our percentage of 30% (three out of ten patients) who chose the BAHA is in accordance with these reports

In conclusion

Most of the patients experienced some degree of benefit with each of the three CROS devices, but it was not large enough to outweigh the disadvantages Preference for one of the three hearing aids was independent of the order in which they were tried We recommend that all patients with unilateral inner ear deafness who apply for hearing aids should be offered a trial with at least the BAHA CROS on a headband It would be worthwhile to formulate selection criteria to help establish which hearing aid will provide the most benefit

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5

General discussion

General Discussion

During 30 years of clinical experience with the BAHA, it has become a well-established treatment for patients with conductive or mixed hearing loss. Owing to the good clinical outcomes, use of the BAHA has spread and the indications for application have gradually been widened. Since the BAHA was first introduced in 1988 at the Radboud University Nijmegen Medical Centre, several new indications have appeared, especially in relation with unilateral hearing impairment. Gradually, after good outcomes with the conventional indications for BAHA application, the Nijmegen BAHA team has extended their inclusion criteria. Clinical and patient outcomes have been evaluated and described in several Nijmegen BAHA – PhD theses. In this thesis, one of the issues we addresses was the application of the BAHA in patients with moderate mental retardation and severe unilateral congenital conductive hearing loss. In addition, we analysed the outcome of BAHA CROS application in a large series of 56 patients with unilateral total inner ear deafness. The most recent study comprised a randomized controlled trial in 10 patients to measure their performance with the conventional CROS, the completely in the canal (CIC) and the BAHA CROS.

Moderate mental retardation

Chapter 2 starts with an evaluation of 22 patients with moderate mental retardation and conductive hearing loss. The clinical and audiological outcomes are presented in *chapter 2.1*. Initially, patients with moderate mental retardation were excluded from BAHA treatment. Our study showed that the BAHA produced satisfactory results in such patients. Their exclusion was based on the suspicion that the extrusion rate of the titanium percutaneous implant would be much higher in these patients. However, the BAHA was well-accepted and they used it for most of the day. The extrusion rate seemed to be comparable with other groups of BAHA users. Although implant loss was low, extra attention may be required from the patients' care providers to preserve the percutaneous implant. Implementation of the BAHA proved to be sufficiently effective. With the BAHA, mean free–field thresholds showed a clear mean improvement of 9 dB compared to the previous hearing aid Considerable improvements in daily activities were seen in at least five patients.

The impact and subjective benefit of BAHA application in patients with moderate mental retardation and mixed or conductive haring loss are described in *chapter 2.2*. The GCBI showed that the BAHA was beneficial in this special group of patients. These findings were comparable with those in the control group and

consistent with the results of earlier studies that used the GCBI The LIFE showed overall improvement in listening and learning capabilities This was reflected in considerable positive effects on activities at school or at work As patients with moderate mental retardation are prone to difficulties in development and social communication, it is essential to maximize their hearing, so that social communication is not compromised even further Problems with recurrent ear infections caused by conventional hearing aid fitting can be overcome by the BAHA Extension of the indications for BAHA application to this special patient group proved to be a very good decision

Unilateral congenital conductive hearing loss

In chapter 3, we report the audiometric results (chapter 3 1) and the subjective benefit (chapter 3 2) of the BAHA in a consecutive group of 20 patients with severe unilateral congenital conductive hearing impairment The results were compared to those obtained from a previous consecutive group with severe unilateral acquired conductive hearing loss (Hol et al 2005) [1]

It is still a challenge to achieve binaural hearing in patients with unilateral congenital hearing impairment Very few patients with unilateral congenital ear canal atresia are suitable candidates for microsurgery Moreover, in children with ear canal atresia, surgical intervention is usually postponed until the age of 6 years An alternative to surgery is to fit a conventional air conduction hearing aid However, in patients with complete bony ear canal atresia, this is not an option Furthermore, in cases with a large air-bone gap, very high levels of amplification are needed with an air conduction hearing aid, which cannot always be provided owing to feedback problems A bone conduction hearing aid, such as the BAHA system, seems to be a better option In patients with unilateral acquired conductive hearing loss, the BAHA successfully restored binaural hearing Hol et al reported significantly improved sound localization, speech recognition and subjective benefit in 18 patients with a unilateral acquired air-bone gap [1] In an earlier pilot study, Snik et al (2002) reported that although the BAHA effectively improved directional hearing in the patients with unilateral acquired conductive hearing loss, the results were ambiguous in the two patients with unilateral congenital conductive hearing impairment[2] Directional hearing tests, after BAHA fitting, did not show any improvement in the latter two patients, because their localization abilities were already good in the unaided condition

Over a period of 8 years, 20 patients with unilateral congenital conductive hearing impairment have been fitted with a BAHA In 6 out of the 18 patients with a complete data set, the results of sound localization tests did not show any

significant improvement Speech recognition in noise with spatially separated speech and noise sources improved after BAHA application, but not significantly There was remarkably close compliance with BAHA use in this patient group, which suggests high patient satisfaction The subjective benefit evaluation (chapter 3 2) on the 10 adults and 10 children showed that the BAHA was well-accepted by most of those patients Nowadays, a preoperative trial with the BAHA on a steel headband is part of the routine preoperative procedure

Early intervention is still not considered to be indicated in children with unilateral ear canal atresia, provided that the child has normal hearing in the other ear It is rarely recommended to fit a conventional air conduction or bone conduction hearing aid, because many authors found poor acceptance by most children In contradiction, recent clinical experience has shown that after successful surgery, children with bilateral severe congenital conductive hearing loss due to isolated ossicular chain anomalies started to use their hearing aid in the non-operated ear once the other ear had started to hear well Based on our own experience, it is important to consider treatment with a hearing aid when children with unilateral (congenital) severe conductive hearing impairment are found to have delayed speech and language development The new BAHA-softband provides a unique trial opportunity for young children with unilateral congenital ear canal atresia Our results in patients with unilateral congenital conductive hearing loss confirmed the subjective benefit of the BAHA recently reported by Priwin et al in a small group of patients [3] A preoperative trial with a transcutaneous BAHA is advisable before a decision is made More detailed evaluation of patient characteristics might reveal additional relevant criteria to help indentify the best candidates for BAHA treatment Application of the BAHA in this group of young patients will provide completely new opportunities to study the mechanisms of bone conduction and central hearing in the near future

BAHA CROS application in patients with single sided deafness

Chapter 4 1 describes the use of the BAHA by patients with unilateral profound sensorineural hearing loss The detrimental effects of unilateral hearing loss are more fully recognized nowadays Traditionally, these patients were fitted with a contralateral routing of sound (CROS) hearing aid Due to the small interaural attenuation of sound conducted by bone, the BAHA can be used as a transcranial Contralateral Routing Of Sound (CROS) device

Step by step the outcome of BAHA CROS application has been presented in the consecutive Nijmegen series Other centres have also shown promising results with this new BAHA indication.[4-10] Transcranial BAHA CROS comprises a hearing

aid and microphone on the impaired side, without any occlusion of the external ear canal on the good side

Our study included 28 patients with unilateral acquired and 2 patients with unilateral congenital inner ear deafness During the evaluation, 3 patients dropped-out A total of 56 patients were evaluated, including an earlier series of 29 patients Sound localization results in a well-structured test setting were not above chance level Patients reported some improvement in their capacity to localize sounds with the BAHA CROS in daily life The main advantage of the BAHA CROS was to lift the head shadow effect during the speech in noise test Mean day-to-day use of the BAHA CROS in the total group was 84% All the instruments on subjective benefit with the BAHA CROS showed positive results Therefore, BAHA CROS application seems to be having consistent, positive effects in the Nijmegen series

The promising results with the BAHA CROS may have been influenced by the etiology of deafness, the duration of the total unilateral inner ear deafness and selection procedures Hol et al reported that 9 out of the 39 patients (about 25%) declined BAHA CROS application after a preoperative trial with the BAHA CROS on a headband [9] In patients with unilateral total inner ear deafness as result of acoustic neuroma or its surgical treatment, Andersen et al (2006) reported that only 25% applied for a BAHA CROS after a trial with transcutaneous BAHA CROS application [11] It would be interesting to evaluate the personal and social characteristics of the patients with total unilateral inner ear deafness who benefit from BAHA CROS application and apply for implantation In the recent study by Andersen et al 59 patients with unilateral hearing loss after acoustic neuroma surgery were sent a written invitation for a trial period with the BAHA CROS on a headband [11] Only half of them accepted and only 25% of the participants ultimately opted for the device The outcome of our small (n=10) randomized controlled trial was in agreement with this In the trial, 3 different hearing aids were compared the conventional CROS, the CIC and the BAHA CROS Three out of the 10 patients (30%) chose the BAHA CROS and have been implanted They reported that they are satisfied One patient (10%) chose the conventional CROS with an FM link instead of a wire around the neck

We recommended that all patients with unilateral total inner ear deafness who apply for hearing aids should be offered a trial with the conventional CROS and the transcutaneous BAHA CROS

In our series of 56 patients with unilateral total inner ear deafness, it was remarkable that only 5 had a congenital etiology This may be partly due to selection, because at the start of our BAHA CROS application patients who had

undergone acoustic neuroma surgery were offered a transcutaneous trial. We also soon found that patients with a congenital etiology were not so interested in this amplification method. In this thesis, we report the outcome of the BAHA CROS in 5 patients with a congenital etiology. It would be worthwhile to compare the outcome of BAHA CROS application in large groups of acquired and congenital patients and to analyse the patient characteristics.

In summary, the BAHA is a valuable treatment option in patients with many different types of hearing loss. The first conclusion in this thesis is that the BAHA can be applied effectively to patients with moderate mental retardation. The second conclusion is that extension of the indications to unilateral severe conductive hearing loss was worthwhile, but also gave rise to some new questions. The third conclusion is that in patients with total unilateral inner ear deafness, the BAHA CROS was only successful in well-selected cases.
Extension of the indications to patients with unilateral congenital severe conductive hearing impairment produced promising results. There were differences in outcome between the unilateral acquired group and the unilateral congenital group. The unilateral congenital patients were found to have very good sound localization abilities prior to BAHA application. These remarkable findings need to be confirmed in new studies. Exploration of the underlying features is an important topic for more fundamental studies on the mechanisms of bone conduction and central hearing function.
There are indications that differences exist in the outcome of BAHA CROS application between patients with unilateral acquired total inner ear deafness and patients with unilateral congenital inner ear deafness. This once again offers opportunities for research into the central pathways of hearing. We also need to understand which determinants will lead to better performance and greater patient satisfaction after BAHA CROS application.
Since the introduction of the BAHA 30 years ago, indications for its clinical application are still expanding and opening new fields for research, not only into the mechanism of hearing via bone conduction in general, but also into specific groups of patients, such as those with unilateral congenital total deafness. BAHA application has made very valuable contributions to many hearing impaired patients and can be expected to extend to new groups in the future.

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6

Summary and Conclusions

During thirty years of clinical experience, the bone anchored hearing aid (BAHA) has become a well-established treatment for hearing impaired patients with bilateral conductive or mixed hearing loss Several studies outside this thesis have shown that the BAHA is a highly effective treatment in patients with these conventional indications

Since the BAHA was first introduced at our centre in 1988, a number of new indications have been formulated Good outcomes with the conventional indications have gradually led to extension of the inclusion criteria by the Nijmegen BAHA team Clinical and patient outcomes have been analysed in 4 previous theses Recent research, including the studies described in this thesis, focused on the new extended indications

Initially, patients with moderate mental retardation were excluded from BAHA treatment As these patients are likely to encounter difficulties with development and social communication, adequate treatment with hearing aids is essential to ensure that their social communication is not compromised even further In 22 patients with moderate mental retardation and conductive hearing loss, audiometric and subjective evaluations after BAHA fitting showed encouraging results The BAHA was well-accepted by these patients and they were using it for most of the day Implant extrusion rates seemed to be comparable with those of the general BAHA user population

Another new BAHA indication is congenital unilateral conductive hearing loss Earlier evaluation of BAHA patients with severe acquired unilateral conductive hearing loss by Hol et al (2005) revealed very promising outcomes In this thesis, patients with congenital unilateral conductive hearing loss were evaluated The BAHA was well-accepted by most of the patients with congenital unilateral conductive hearing impairment Some patients had such good directional hearing and speech-in-noise scores in the unaided situation that no overall significant improvement occurred after BAHA fitting in our set-up However, observations of consistent use of the device were highly suggestive of patient benefit Further research is recommended to gain more insight into these findings

The BAHA system placed on the shadow side of the head proved advantageous in patients with unilateral inner ear deafness, due to contralateral routing of sound via bone conduction However, as these patients have only one functioning cochlea, their sound localization abilities did not improve in our experimental set-up Patient outcome instruments showed encouraging and consistent results in an extended group of 56 patients In a randomised controlled trial, 10 adult patients with unilateral inner ear deafness tested three different CROS hearing aids the BAHA CROS, the conventional CROS and the completely-in-the canal hearing aid

(CIC) Patients experienced benefit with each of the three hearing aids and their outcome maesurements showed improvement, particularly with the BAHA CROS Therefore, in patients with unilateral inner ear deafness, an evaluation to find out which hearing aid suits them best, based on their needs and expectations, could be of additional value

Nowadays, the BAHA is a valuable treatment option in patients with many different audiological problems and comorbid conditions We found that the BAHA could be used effectively by patients with moderate mental retardation and conductive hearing loss In children with unilateral conductive hearing loss, the BAHA can be considered as an option to assist development and overcome communication difficulties The BAHA has also proved to be a consistent treatment in patients with unilateral inner ear deafness Nevertheless, additional audiological and patient outcome measurements are needed to provide further support for these new indications With the introduction of partially implantable hearing devices or middle ear implantations, the field opens to discuss strategies to choose between the different rehabilitation options It is important to continue to evaluate the BAHA to demonstrate its value and to consolidate its strong position among all the other hearing rehabilitation methods However, during thirty years of experience, the BAHA has proved to be a consistent and valuable option for hearing impaired patients

Na dertig jaar klinische ervaring is het in het been verankerd hoortoestel (BAHA) een bewezen revalidatie methode voor patienten met een bilateraal geleidings of gemengd gehoorverlies Verscheidene studies buiten dit proefschrift hebben aangetoond dat het BAHA systeem een effectieve behandeling is voor patienten met deze conventionele indicaties Sinds de introductie van de BAHA in het Universitair Medisch Centrum Nijmegen (UMCN) in 1988 zijn een aantal nieuwe indicaties toegevoegd Goede resultaten met de conventionele indicaties leidden geleidelijk aan tot uitbreiding van de inclusie criteria door het BAHA team in Nijmegen Een evaluatie van klinische resultaten en patient tevredenheid resulteerde in 4 voorgaande proefschriften Recente onderzoeken, onder andere in dit proefschrift beschreven, onderbouwen een verdere uitbreiding van de indicaties

Patienten met een milde mentale retardatie en een geleidings gehoorverlies werden aanvankelijk uitgesloten van behandeling met een BAHA Aangezien deze patienten eerder ontwikkelings en communicatie problemen kunnen ontwikkelen, is een adequate gehoorrevalidatie essentieel om een zo goed mogelijk gehoor te waarborgen, zodat de sociale communicatie niet nog verder wordt gecompromitteerd De audiometrische en subjectieve evaluatie van 22 patienten met milde mentale retardatie en een geleidings gehoorverlies liet na aanpassing met een BAHA positieve resultaten zien De BAHA werd door deze patienten goed geaccepteerd en voor het grootste deel van de dag gedragen Het percentage van schroefverlies was vergelijkbaar met dat van BAHA gebruikers met conventionele indicaties

Een andere nieuwe BAHA indicatie is revalidatie van patienten met een eenzijdig aangeboren geleidingsverlies Een evaluatie van Hol et al (2005) naar toepassing van de BAHA bij patienten met een eenzijdig verworven geleidingsverlies toonde hoopgevende resultaten In dit proefschrift werden patienten met een eenzijdig aangeboren geleidingsverlies geevalueerd. De BAHA werd in deze groep goed geaccepteerd door de meeste patienten Tijdens het testen van richtinghoren konden sommige patienten bij de ongeholpen metingen al zo goed geluid lokaliseren dat er bij de geholpen situatie (met BAHA) geen significante verbetering werd gevonden in onze testsituatie Daarentegen werd de BAHA consistent gedragen wat suggestief is voor een goede patient tevredenheid Verder onderzoek is nodig om deze resultaten meer inzichtelijk te maken.

Patienten met een eenzijdige binnenoordoofheid hebben baat bij een BAHA die aan de schaduwzijde (dove kant) van het hoofd geplaatst wordt, waarbij geluid via beengeleiding naar de contralaterale zijde (goedhorende kant) geleid wordt Aangezien deze patienten slechts een functionerend slakkenhuis hebben, verbeteren zoals verwacht de vaardigheden om geluid te lokaliseren niet in onze testsituatie Evaluatie naar patient tevredenheid toonde bemoedigende en vergelijkbaar consistente resultaten aan in de patientengroep met eenzijdige binnenoordoofheid, uitgebreid naar 56 patienten
In een prospectieve studie van 10 patienten met een eenzijdige binnenoordoofheid werden drie verschillende methodes van gehoorrevalidatie geevalueerd Naast de BAHA CROS en de conventionele CROS werd ook een volledig in het oor hoortoestel (CIC) geevalueerd Van elk van de methodes ondervonden de patienten voordeel Patient tevredenheid kwam het meest naar voren met de BAHA Bij patienten met eenzijdige binnenoordoofheid kan een evaluatie van behoeftes en verwachtingen van de patient van aanvullende waarde zijn voor de keuze van het te gebruiken systeem

Het BAHA systeem is tegenwoordig een waardevolle behandelingsoptie voor veel verschillende patientengroepen Uit dit proefschrift komt naar voren dat de BAHA effectief is voor patienten met een milde mentale retardatie en een conductief gehoorverlies Bij kinderen met een eenzijdig conductief verlies, waarbij ontwikkelings- en communicatieproblemen kunnen ontstaan, kan interventie met een BAHA overwogen worden Patienten met eenzijdige binnenoordoofheid bleken consistent baat te hebben bij behandeling met het BAHA systeem Extra audiologische metingen en subjectieve evaluaties zijn nodig om deze nieuwe indicaties verder te onderbouwen
De introductie van nieuwe technieken op het gebied van middenoor implantaten opent de discussie over de keuze tussen de verschillende revalidatie methodes Evaluatie van de BAHA in de toekomst blijft belangrijk om de waarde en de plaats van de BAHA binnen deze nieuwe ontwikkelingen te bepalen Echter op dit moment is de BAHA, na dertig jaar een consistente en waardevolle optie gebleken binnen de gehoorrevalidatie

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Dankwoord
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Dankwoord

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Sylvia Kunst, april 2008

Curriculum Vitae

Sylvia J W Kunst werd geboren op 18 februari 1979 te Breda In 1997 behaalde zij haar Gymnasium diploma aan het St Oelbert Gymnasium te Oosterhout (NB) In datzelfde jaar begon zij aan haar studententijd in Nijmegen Na uitloting voor de studie Geneeskunde koos zij voor Biologie aan de Radboud Universiteit In 1998 behaalde zij haar propedeuse Biologie en tegelijkertijd kon ze beginnen aan de opleiding Geneeskunde eveneens aan de Radboud Universiteit Het doctoraal-examen werd behaald in 2002 Na een wetenschappelijke stage onder leiding van Prof dr H A M Marres naar het nasopharynxcarcinoom werd in 2003 gestart met haar co-schappen Tijdens een keuze co-schap Keel-, Neus- en Oorheelkunde werd reeds gestart met een wetenschappelijk onderzoek naar de toepassing van de BAHA bij milde mentaal geretardeerde kinderen in samenwerking met Mevr dr M K S Hol In november 2004 behaalde zij haar artsexamen Aansluitend werd zij aangesteld als arts-onderzoeker bij de afdeling Keel-, Neus- en Oorheelkunde van het UMC St Radboud Onder leiding van Prof dr C W R J Cremers werkte zij mee aan een groot Europees onderzoek naar ouderdom slechthorendheid, het ARHI project In 2005 werd dit project succesvol afgerond Hierna kon de verdere basis worden gelegd voor het wetenschappelijk onderzoek beschreven in dit proefschrift naar de uitbreiding van indicaties voor de BAHA Hierin werd zij begeleid door het Nijmegen BAHA-team onder leiding van Prof dr C W R J Cremers In juli 2006 startte zij met haar opleiding tot medisch specialist in de Keel-, Neus-, en Oorheelkunde (hoofd opleider Prof dr K Graamans) In het Canisius Wilhelmina Ziekenhuis te Nijmegen (opleider Dr J A M Engel) en in het VieCuri Medisch Centrum te Venlo (opleider Dr E Theunissen) doorliep zij de afgelopen maanden haar perifere opleidingsstages

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AD	Auricula dextra
AC	Air conduction
ACHA	Air conduction hearing aid
APHAB	Abbreviated profile of hearing aid benefit
AS	Auricula sinistra
AV	Aversiveness of sound (domain of APHAB)
BAHA	Bone anchored hearing aid
BC	Bone conduction
BCHA	Bone conduction hearing aid
BE	Best ear
BN	Background noise (domain of APHAB)
BCHA	Bone conduction hearing aid
CIC	Completely in the canal
CROS	Contralateral routing of sound
dB	Decibel
dBnHL	Decibel relative to normal hearing level
DCC	Day care centre
Deg	Degree
EC	Ease of communication (domain of APHAB)
Eci	E causa ignota
FF	Free field
FM	Frequency modulation
GBI	Glasgow benefit inventory
GCBI	Glasgow children benefit inventory
GHABP	Glasgow hearing aid benefit profile
HHDI	Hearing handicap and disability inventory
HL	Hearing level
Hz	Herz
IOI-HA	Intern Outcome Inventory for Hearing Aids
LIFE	Listening inventory for education
MAE	Mean absolute error
MPS	Maximum phoneme score
NH	Normal hearing side
PE	Poor ear
Pinna	Auricle
PTA	Pure tone average (mean hearing loss at 0.5, 1, 2 and 4 kHz)
RV	Reverberation (domain of APHAB)
S/N ratio	Speech-to-noise ratio
SPL	Sound pressure level (decibel)
SRT	Speech reception threshold
SSD	Single sided deafness
SSQ	Speech spatial and qualities of hearing scale


BAHA!