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BONE ANCHORED HEARING AIDS (BAHAs) IN CHILDREN

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ABSTRACT

BACKGROUND: The technique for applying bone-anchored hearing aids (BAHAs) by the use of osseointegration is today widely established for both adults and children. The BAHA concept is suitable for patients with recurrent ear infections or ear malformations who cannot use conventional hearing aids, which are placed altogether or partially in the ear canal. Long-term results are well documented in adults but are lacking in children. Most patients are fitted with unilateral BAHA even though they have bilateral hearing loss. However, recent studies in adults with bilateral hearing loss have shown better hearing results with bilateral BAHA fitting compared to unilateral. Moreover, a recently published metaanalysis on the consequences of unilateral hearing loss in children has implied that educational and behavioral problems as well as delayed speech and language development can occur. Improved hearing outcomes have lately been presented in adults with single-sided deafness and unilateral conductive hearing loss fitted with unilateral BAHA why unilateral BAHA ought to be explored also in children.
AIMS: The study was performed with the aim to increase our knowledge of long-term results and new areas of use of BAHA in children with either uni- or bilateral conductive hearing loss. A further objective was to evaluate the influence of single-sided congenital conductive hearing loss on hearing function and to investigate perceived problems in patients with congenital ear malformation where no hearing intervention has been performed.
MATERIAL AND METHODS: Altogether 127 patients and 15 controls, mainly children were enrolled in the present study. The included subjects had either uni-or bilateral conductive hearing loss. The patients with bilateral hearing loss were fitted with BAHAs uni-or bilaterally whereas the patients with unilateral hearing loss were predominantly untreated or fitted with unilateral BAHA. Medical records were examined. Psychoacoustic tests including tone and speech audiometry, sound localizations tasks, as well as satisfactory measurements were employed.
RESULTS AND CONCLUSION: Long-term results in children concerning implant failure rate, adverse skin reactions and function pattern of the BAHA were in parity with previous measured outcomes in adults. Furthermore, bilateral BAHAs in patients, both children and adults gave additional hearing effects such as improved speech reception/recognition and sound localization ability as well as binaural hearing to some extent. Unilateral hearing aid fitting in children gave some supplementary benefit in terms of improved speech recognition in noise but no positive influence on sound localization ability was recorded. Nevertheless, all children fitted with hearing aids, either uni-or bilaterally reported positive outcome with their devices. Two problem areas were identified in the hearing impaired children: in reaction to sounds and with speech intelligibility. Finally, in patients with single-sided congenital external ear malformations and associated hearing loss, a deprived auditory function was noticed. This patient cohort also reported a high degree of hearing related problems.
In conclusion, BAHA is a good amplification alternative in children with conductive hearing loss. Bilateral BAHAs resulted in supplementary hearing gain compared to one BAHA in both adults and children why bilateral BAHAs could be considered in patients with bilateral conductive hearing loss. Unilateral hearing aid, mainly BAHA might also be beneficial to some extent in children with conductive unilateral hearing loss and a trial with BAHA on Softband could be considered. Complementary intervention should also focus on the alleviation of the high degree of self and guardian reported problems in audition and communication.

Keywords: BAHA, conductive hearing loss, unilateral hearing loss, children, audiometry, sound localization, speech audiometry, self-assessment, questionnaire, craniofacial abnormalities

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LIST OF PUBLICATIONS

This thesis is based on the following original papers, they will be referred to in the text by their Roman numerals I – IV.

I. Priwin C, Granström G. The bone-anchored hearing aid in children: a surgical and questionnaire follow-up study. *Otolaryngol Head Neck Surg* 2005; 132: 559-65

II. Priwin C, Stenfelt S, Granström G, Tjellström A, Håkansson B. Bilateral bone-anchored hearing aids (BAHAs): an audiometric evaluation. *Laryngoscope* 2004; 114: 77-84

III. Priwin C, Jönsson R, Hultcrantz M, Granström G. BAHA in children and adolescents with uni- or bilateral conductive hearing loss. A pilot study of outcomes. Submitted for publication.

IV. Priwin C, Jönsson R, Magnusson L, Hultcrantz M, Granström G. Audiological evaluation and self-assessed hearing problems in subjects with single-sided congenital external ear malformations and conductive hearing loss. Submitted for publication.

CONTENTS

LIST OF ABBREVIATIONS

AC	air conduction
ACHA	air conduction hearing aid
BAE	bone anchored episthesis
BAHA	bone anchored hearing aid
BC	bone conduction
BCHA	bone conduction hearing aid
BHL	bilateral hearing loss
BMLD	binaural masking level difference
CBC	conventional bone conductor
CROS	contralateral routing of signal
CSOM	chronic suppurative otitis media
dB	decibel
DBC	direct bone conduction
EM	ear malformation
EO	external otitis
HA	hearing aid
HL	hearing level
ILD	interaural differences in sound level
IOI-HA	International Outcome Inventory for Hearing Aids
ITD	interaural differences in time
kHz	kiloHertz
MAIS	meaningful auditory integration scale
MUSS	meaningful use of speech scale
PB	phonemically balanced
PTA	pure tone average
PTA (M3)	pure tone average of thresholds at 0.5,1 and 2 kHz
PTA (M4)	pure tone average of thresholds at 0.5, 1, 2 and 4 kHz
SBU	Swedish Council of Technology Assessment in Health Care
SD	standard deviation
SII	speech intelligibility index
S/N ratio	signal-to-noise ratio
SPBN	Swedish phonemically balanced words in noise
SPL	sound pressure level
UHL	unilateral hearing loss

1 INTRODUCTION

In 1977, a new technique to establish bone-anchored hearing aids (BAHAs) by use of osseointegration implants was developed in Gothenburg, Sweden [1,2]. The aim was to improve rehabilitation for patients with conductive or mixed hearing loss and in addition to offer an optional hearing aid (HA) for patients unable to wear conventional air conductive hearing aids (ACHAs).

The BAHA is today the only HA system available, which applies the use of direct bone conduction (DBC). The device consists of a vibrating sound processor attached to the temporal or parietal bone by an osseointegrated implant. In such way, sound transmission from the transducer to the skull base, where the cochleae are situated takes place with no interference from the skin and soft tissue [3].

By the use of DBC, several problems with both conventional air and bone conductive devices can be eliminated. BAHA is, for instance, an alternative for patients with moderate sensorineural hearing loss who do not tolerate an ACHA placed partially or altogether in the ear canal due to recurrent ear canal infections or chronic suppurative otitis media (CSOM). Further, the sole HA option in patients with external EM causing conductive hearing loss used to be conventional bone conductors (CBC). The CBC has major drawbacks such as poor sound quality and poor aesthetics as well as a tendency to generate pain over the mastoid area due to the high pressure needed to maintain adequate contact between the device and the bone. In these patient cohorts, BAHA has to large extent replaced CBC in many countries.

According to the manufacturer's data, (Entific, Sweden, 2005) more than 25 000 patients have been fitted with BAHA worldwide since its introduction and the number is rapidly increasing. Most fitted patients are adults but the BAHA concept is since a long period approved also in children. Numerous studies have been presented by the different BAHA centers concerning surgery, audiological outcomes and patient benefits with BAHA in both adults [4, 5, 6, 7, 8, 9] and children [10, 11, 12, 13, 14, 15, 16].

1.1 HEARING IMPAIRMENT

1.1.1 General aspects

Hearing is of utmost importance in our ability to communicate with other people. A hearing impairment will debase communication skills and thus reduce the capacity to interact with the surrounding community. In other words, a hearing handicap might result in social isolation for the affected individual. Further, if hearing impairment is established early in life, speech and language development might also be suppressed with even more devastating consequences [17].

1.1.2 Prevalence

The prevalence of hearing impairment is rather high in the general population. 10.7 % of Swedish inhabitants between 16-84 years of age reported self-assessed hearing problems in noisy surroundings covered in a study conducted by Rosenhall in 1997 [18].

With increasing age (> 50 years) hearing capacity subsides further and hearing loss is even more frequent. In a survey of healthy 70-year old Swedish inhabitants, the corresponding number complaining of hearing loss was 55-60% [19].

Altogether 1.3 million individuals >18 years of age suffered from slight to severe hearing impairment in Sweden 2002 of whom approximately 560 000 would benefit from HA fitting according to the Swedish Council of Technology Assessment in Health Care, SBU (www.sbu.su).
Approximately 50 % of the individuals with a hearing handicap that would benefit from hearing amplification had received hearing rehabilitation measures with HA fitting. Nevertheless, hearing rehabilitation within the community was expensive and the total cost of HA fitting in Sweden year 2002 was 562 million Swedish crowns where the actual HA cost was 287 million Swedish crowns according to SBU.

1.1.3 Optional treatments

Audiological rehabilitation has been described as a patient orientated process driven by the difficulties that the individuals and those around perceive. A definition proposed by Stephens [20] is "a problem solving process aiming to minimize the disablements experienced by individuals with hearing disorders and to maximize their quality of life".

At present, optional treatments for hearing impairment consist of either reconstructive ear surgery or technical aids. Technical aids include HAs, reinforced doorbells, telephones, alarm clocks etc. There are many different types of HAs and which one is suitable on an individual basis depends on the degree and type of hearing loss. The most common type of HA is the ACHA, which uses either analogue or digital sound processing technique [21]. Other types of hearing devices consist of cochlear implants used in profoundly deaf patients and BAHAs suitable for patients with conductive type of hearing loss or in patients with recurrent ear infections unable to use conventional HAs [22].

Currently, the prevailing treatment of hearing loss is HA fitting since it suits all kinds of hearing impairments regardless of etiology and type i.e. conductive or sensorineural hearing loss. Ear surgery is an exclusive treatment alternative in conductive hearing loss caused by middle ear pathology such as otosclerosis and CSOM. Furthermore, reconstructive ear surgery is also a treatment option in patients with external EMs, for example ear canal atresia, though the results are ambiguous and depend on the extent of the malformation [23]. Until today, no surgical techniques are known to be developed in the clinical scenery to repair or exchange damaged hair cells in the inner ear or to interact with the central auditory system with for instance stem cells. Intensive research is ongoing in the area of stem cell implantation and promising results have so far been presented in animal models [24].

Until recently, mainly patients with bilateral hearing impairment were considered as candidates for general hearing intervention. However, an exception has always been treatment of single-sided hearing loss due to middle ear disease where surgical treatment is possible. Other cases with unilateral hearing loss (UHL) have remained without rehabilitation since good hearing in one ear has been considered satisfactory [25]. However, a recent published metaanalysis on the consequences of UHL in children showed an increased number of educational and behavioral tribulations among the children with an increased rate of grade failures as well as excessive need of educational assistance compared to otologically normal children. It was also concluded that UHL could cause delayed speech and language development and it remained unclear if the children caught up with their normal hearing peers if treated [25].

Hence, hearing handicaps are common throughout the society and rehabilitation efforts remain costly. All the same, hearing loss, either uni- or bilaterally might well result in vastly harmful consequences on both individual and socioeconomic basis why it is highly central to offer good rehabilitation alternatives for all hearing impaired subgroups. Patient groups benefiting from BAHA are limited but nevertheless essential.

1.2 CHARACTERISTICS OF BONE CONDUCTION

1.2.1 Physiology

Hearing by bone conduction (BC) is a far more complex function than hearing by the ordinary air conductive route of sound waves. Although the phenomenon of hearing in sentence of vibrations in the skull has been known for more than a century, yet its physiology has not been fully understood despite intensive and thorough research in this field.

It was early acknowledged that both air conduction (AC) and BC stimulation result in similar basilar membrane motion in the cochlea [26]. This motion is coded by the inner ear neurons and then interpreted in the primary and secondary auditory cortices as sounds.

In a more recent study, five different factors contributing to BC hearing have been identified [27]:

1. Sound radiated into the external ear canal
2. Inertia of the ossicles in the middle ear
3. Inertia of the cochlear fluids
4. Compression of the cochlear wall
5. Transmission of pressure from the cerebrospinal fluid

Of the mentioned factors, the overall most important contributor to BC hearing in the normal ear seems to be the inertia of the cochlear fluid, especially in the lower frequency range. The importance of the different factors is, however, highly frequency dependent and for instance the inertia of the middle ear ossicles is important in the mid frequencies (1-3 kHz) and sound radiated into the external ear canal is of importance in the low frequencies if the ear canal is blocked [27].

1.2.2 Transcranial attenuation

A frequency dependent response difference to vibrations between the two cochleae is termed transcranial attenuation. The subjective transcranial attenuation was found to be approximately 10 dB for the frequencies 0.25 to 4 kHz with great individual difference [28]. Furthermore, measurements conducted by Stenfelt & Håkansson [29] on dry skulls added with dampening material showed higher responses on the contralateral side in the lower frequency range (0.5-1 kHz) due to an anti-resonance effect. In the higher frequencies, above 1 kHz, the response level was higher at the ipsilateral side. The transcranial attenuation varied in this experiment between –5-10 dB.

1.2.3 Clinical usage of the BC route

Knowledge about the BC transmission route of sound waves applies for two different areas in the clinical setting.

1.2.3.1 Diagnostics

Testing the BC ability has mainly been used in the past decades to differentiate sensorineural and conductive hearing impairment where a discrepancy between the AC and BC thresholds, i.e. an air-bone gap indicates a conductive type of hearing loss. Furthermore, BC sound is considered to reflect true cochlear function why the AC and BC thresholds should not differ in normal hearing or in a sensorineural hearing loss where no interruption of the sound wave's way to the inner ear is present [27].

The ability to test the BC route of sound waves, by generating a vibration of the temporal bone is essential and can be achieved in three different ways:

1. Percutaneous stimulation of the temporal bone (BAHA)
2. Transcutaneous stimulation through the skin (Radioear B-71 bone transducer, tuning forks)
3. Induced vibration from AC sound (warble tones in a sound field)

Generally, BC is tested with pure tones in the frequency range 0.25 to 4 kHz where a bone transducer is placed over the mastoid area. In the outpatient clinic, a simpler way to differentiate between sensorineural and conductive hearing loss is the use of tuning fork tests i.e. Weber, Rinné and Schwabach test. However, BC can also be tested with more modern techniques such as auditory brainstem response (ABR) [27].

1.2.3.2 Hearing rehabilitation

BC is known to be a less efficient way of hearing than AC why ACHAs are mainly used in hearing rehabilitation and bone conductive hearing aids (BCHA) are merely used when contraindications for ACHAs are present.
Thus, patients with conductive hearing loss or sensorineural hearing loss unable to wear an ear mould in the ear canal are suitable candidates for the BC route of sounds. At present, there are two fundamentally different types of BCHAs available on the market i.e. CBC and BAHA.

1.3 OSSEOINTEGRATION

1.3.1 Background

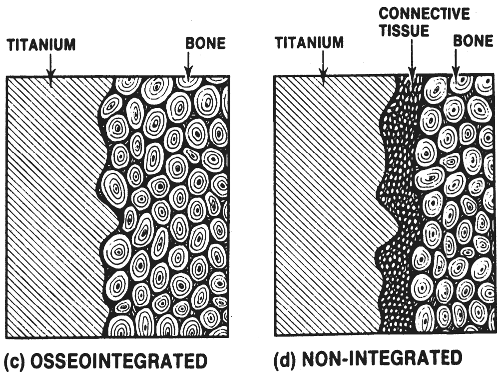
The osseointegration concept was developed by Per-Ingvar Brånemark in Sweden during the late 50s and 60s. He primarily defined osseointegration in theory and

practice [30] as “A direct structural and functional connection between ordered, living bone and the surface of load-carrying implant “.
Initially, his experimental work was focused on observing microcirculation in bone marrow. During the experiments, a modified vital microscopi chamber was used, where a titanium implant with a central canal and a transverse opening at one level was threaded into bone to allow the growth of bone and vessels into the chamber. The titanium chambers showed a high tendency to become permanently incorporated with the bone why the possibilities of integrating titanium screws into bone came into focus and intensive research was immediately commenced in this area [31].

Historically, numerous materials have been tested in the field of bone anchored implants [32]. At first, various metallic implants were attempted due to their outstanding mechanical properties. Unfortunately most metals, except non-alloyed titanium, vitallium and tantalum, resulted in development of a fibrous tissue coating around the implant as a response to the foreign material and thus no osseointegration took place [32, 33]. Later diverse biocompatible surfaces, for instance bioglass have been tried [32]. These materials might be accepted in the bone without trigging an inflammatory response but have poor mechanical characteristics.
To date, c.p, commercially pure titanium is used in the field of osseointegration due to its unique characteristic to form an oxide layer on the surface which facilitates the process of osseointegration [33].

1.3.2 Principles of osseointegration

Fig I. An illustration of an osseointegrated versus a non-integrated implant. (*The figure is published by permission of Barbro K.Brånemark)*



An implant is nowadays defined as osseointegrated if there is no progressive relative motion between the living bone and the implant under functional levels and types of loading (fig I). Multiple biological factors determine if osseointegration will become successful or not. Parameters of interest for the process are the materials biocompatibility, implant design and structure, status of the recipient bone, surgical approach and finally loading conditions [33].
As previously mentioned, the most common material to use in conjunction with osseointegration is non-alloyed titanium. Under normal circumstances, a thin oxide layer will cover the pure titanium surface when it is exposed to atmospheric conditions. If the titanium implant is subjected to biological tissues an even more extensive oxide

growth will occur and form a hydrated titanium peroxy matrix [31]. The formation of such a matrix is a unique feature of pure titanium and is probably caused by inflammatory cells producing proteolytic enzymes, cytokines, superoxide and hydrogen peroxide. The formed surface layer is one of the key factors to why titanium implants are not only accepted but also integrated within the bone and cannot be removed unless of fracturing the surrounding bone structures.

Furthermore, the design and structure of the implant is of great importance for successful implantation. A cylindrical, threaded implant with a somewhat rough surface is more beneficial than a similar implant with a smooth surface due to better cellular and bone attachment which prevents movement of the implant [32].

Finally, yet another interesting characteristic of osseointegrated implants is the phenomenon called osseoperception. Osseoperception is the term for the patients' ability to identify tactile thresholds transmitted through their prosthesis thus providing better function and comfort than ordinary socket prosthesis [31].

1.3.3 Clinical applications of titanium implants

It is estimated that more than six million patients all over the world have received titanium implants since they were introduced in the clinical field in the 60s and the number of users is rising fast. Applications in the oral surgery field heavily dominate but titanium implants are also used in the area of otorhinolaryngology and orthopedics.

1.3.3.1 Dental

The osseointegration concept is used in dental patients to replace lost teeth and the first implant was inserted in 1965 [31]. The technique can be applied to replace single missing teeth as well as to restore partially or completely edentulous patients by implant supported fixed bridges or removable overdentures with attached frameworks. The long-term results have proved significantly better than conventional prosthodontics [31]. Since its introduction 40 years ago, numerous changes have been made to the osseointegration technique. For instance, surgery is nowadays performed in the outpatient clinic instead of in the operating theatre, the implant itself has been modified and at present different types of coatings as well as optimal roughness of the surface are investigated.

1.3.3.2 BAHA

The first extraoral application of titanium implants was the BAHA where the first three patients were implanted in 1977 [34]. Today, the use of BAHA is well spread over the globe and the BAHA is approved by the food and drug administration, FDA in the United States for use in adults and children > 6 years of age. However, in other parts of the world BAHA is approved also in younger children and the youngest patient in Sweden to receive a BAHA was 18 months of age at the time of the surgery [35].

1.3.3.3 Facial epistheses

Loss of facial tissue due to trauma or tumors is often challenging to reconstruct and at times the results are poor. In complicated cases, implant supported maxillofacial epistheses offer an optional rehabilitation path. The application of the osseointegration concept has been successfully used both in nonirradiated and radiated patients to provide an anchorage for craniofacial epistheses including ears, eyes and noses [31]. Implants in patients with facial deficits are a collaborative task for a surgeon familiar with the procedure and most importantly an anaplastologist. With the use of the implant techniques to secure episthesis, the facial epitheses have become more hygienic and comfortable as well as more satisfactory to the patients than earlier models retained with adhesives.

1.3.3.4 Finger joint prostheses

Another area of use for the osseointegration technique is the replacement of finger joints in patients with rheumatoid arthritis, osteoarthrosis or posttraumatic/postinfection artrosis who often suffer from impaired hand function. Titanium fixtures are then placed in both the phalangeal and the metacarpal bones to support an artificial silicone joint [36].

1.3.3.5 Lower limb prostheses

Lately titanium implants have been used to secure limb prostheses after transfemoral (high) amputation. The results of osseointegrated limb prostheses are promising as an alternative to the traditional socket prostheses, which have complications related to retention and function. The earlier described feature of osseoperception most likely plays a vital roll in the better function seen with the osseointegrated prostheses [31].

1.4 BAHA

1.4.1 Function and fitting range

As previously mentioned, the BAHA works by the route of DBC where vibrations are transmitted percutaneously to an internal titanium implant incorporated in the temporal bone and to both cochleae.
A BAHA consists of an external sound processor carried either at ear-level or body-worn, an abutment and an internal titanium implant. The BAHA housing consists of an electrodynamic transducer with wide frequency range, high output capability, low level of distortion and low levels of current consumption [34].

At present, four different BAHA transducers are available on the market of which three are worn at ear-level (BAHA Classic 300, BAHA Compact and BAHA Divino) and one is body-worn (BAHA Cordelle II). The processors worn at ear-level are suitable for patients with BC PTAs better than 45 dB HL for the frequencies 0.5, 1, 2 and 3 kHz and the corresponding level for the body-worn device is an average threshold better than 60 dB HL according to the manufacturer (www.entific.com).
The ear-level devices close the air-bone gap with a possible additional compensation of roughly 5-10 dB [37]. Furthermore, the BAHA Cordelle's output exceeds the ear-level device, BAHA classic with approximately 15 dB [38].
The latest introduced model is the BAHA Divino where a directional microphone has been incorporated and digital sound processing is used as opposed to the earlier models. In the fall 2006 yet another model, BAHA Power will be introduced on the market

according to the manufacturer. In this device, analog sound processing technique will be used again.

A HA gain characteristic i.e. the quotient between in- and output amplitude is of great importance in choosing the right device for a certain hearing loss. There are two methods of measuring gain, either the insertion gain method or the functional gain method. The insertion gain method is not applicable when measuring gain of a BC device since it is dependent of determining the difference in sound pressure in the ear canal with and without aid. That leaves the functional gain method for BCHA i.e. aided versus unaided thresholds measured in a sound field. In ACHAs the functional gain is determined by the device performance alone but in BCHAs the dignity of the air-bone gap is also of importance. The functional gain is low in patients with air-bone gap close to zero and high in patients with maximal conductive hearing loss, i.e. air-bone gap close to 60 dB. Thus a BAHA becomes relatively better with increasing air-bone gap in comparison to an ACHA [39].

How the wearer will experience the HA is however not only dependent on the gain characteristics but also of the maximum output and maximum dynamic range. The maximum dynamic range is defined as the difference between the DBC threshold and the maximum output of the device, thus the range will diminish in a patient with a sensorineural hearing loss [34].
In clinical practice, a device with high gain and low maximum output will be experienced as stronger in low levels, whereas a device with low gain and high maximum output will be experienced as stronger in the high levels [39].

1.4.2 CBC versus BAHA

Before the BAHA era, patients unable to wear ACHAs, which transmit sound via the external ear canal to the inner ear, were destined to use CBCs in order to amplify hearing.

A CBC consists of an amplifier and a transducer attached to a headband or spectacle frames. The bone transducer is applied with a certain force to the skin covering the mastoid process and transmits sound vibrations transcutaneously to the skull base and the cochleae. The CBC has numerous drawbacks such as variations in speech recognition due to variation in pressure between the transducer and the mastoid, discomfort for the user and poor cosmetic appearance. The high static pressure needed to maintain sufficient contact between the transducer is frequently reported to produce pain, skin irritations and/or headaches. Furthermore, the microphone and the vibrational transducer of a CBC is placed contralaterally why the listening environment becomes unnatural [22, 37].

The introduction of BAHA offered an alternative hearing amplification system to patients not satisfied with the CBC. The main advantages with the new system were the removal of the problematic transcutaneous transducer as well as elimination of the sound attenuating tissue layers between the transducer and the skull [40].

The layer of soft tissue covering the bone in the mastoid area has a damping effect on transmitted sound. Tjellström et al. have previously reported a frequency dependent impedance difference between the skull and soft tissue + skull in the range of 10 to 25 dB. Thus as a result of bypassing the soft tissue with a percutaneous implant, better hearing should be achieved [40, 41].
To verify this theory, multiple studies have compared the audiological and self-assessed effects between the BAHA system and CBC. Enhanced audiological effects in terms of improved free-field thresholds as well as better speech recognition in both quiet and noise have been obtained with BAHA compared to CBC [34, 42, 43, 44, 45, 46]. In addition to the audiological benefits, the patients' overall satisfaction with BAHA were high and BAHA was preferred to CBC by many patients [34, 47, 48, 49]. Furthermore, in a long-term follow-up, conducted by van der Pouw et al., the patients' positive opinion of the speech recognition in quiet and noisy surroundings as well as the quality of sound seemed stable over time [45].

Since one recognized disadvantage with BAHA is the need for a skin-penetrating coupling, a different temporal bone stimulator, the Audiant Bone Conductor was introduced in the mid 80s by Houghs and associates [50]. In this device, Tjellström's technique was modified somewhat and instead of a percutaneous coupling the sound processor was connected to the internally implanted titanium fixture transcutaneously by a magnet, thus skin-penetration was avoided.
However, several comparisons between the two implantable devices have shown clear superiority of the BAHA in terms of better audiological effects even though the transcutaneous device was somewhat more preferable from a cosmetic point of view [51, 52]. The Audiant device has because of its less favourable audiological outcome been removed from the market.

To date, yet another solution is feasible if the skin-penetration site becomes a problem i.e. BAHA mounted on a softband [53]. The softbands are mainly for temporary use in children before surgery is completed but can be utilized on permanent basis if necessary. However, it should be noted that BAHA on softband is less effective than the percutaneous alternative in accordance with any transcutaneous device, why a stronger sound processor might be needed for appropriate hearing amplification [37].

1.4.3 Conventional ACHA versus BAHA

Patients with sensorineural hearing loss are generally equipped with ACHAs since hearing by AC is more effective than hearing with BC. However, approximately 25 % of the patients fitted with ACHAs are dissatisfied with their ACHA and almost half of them use their device less than 2 hours a day [54]. Hence, the need for optional hearing rehabilitation alternatives should always be considered.

In many patients with sensorineural hearing loss, the impairment is most dominant in the high frequency region whilst their low frequency hearing is nearly intact. Applying an ear mould in the ear canal will interrupt the normal sound circuit and consequently it might have a negative influence on their remaining hearing. Furthermore some patients cannot tolerate an ear mould in the ear canal due to various reasons that are more closely displayed in the indication section. With the application of a BAHA, sound is

transmitted to the cochleae via a bypass sound channel thus no adverse effect is to be expected on the remaining hearing, moreover the occlusion effect is avoided and there is less risk of acoustic feedback [55].
A pilot study was performed, where potential benefits of an optimized BAHA for patients with mild to moderate pure sensorineural high frequency impairment were investigated. The results showed better audiological outcomes with an ACHA although the BAHA was preferred for wearing and sound comfort. In conclusion the optimized BAHA used in the study could be a complement to an ACHA but should not be the sole HA in patients with pure sensorineural hearing impairment [55].

Thus a switch from ACHA to BCHA might result in worse hearing outcome; however the results are depending on the presence and dignity of an air-bone gap. In patients using BC device, the ear canal and middle ear is sidestepped, thus the existence and magnitude of an air-bone gap is irrelevant. When using an ACHA on the other side, the device has to compensate for the air-bone gap as well as for any sensorineural component. If the air-bone gap exceeds 25 dB a better outcome with a BCHA could be expected [56].

1.4.4 Indications

The BAHA system is since long a well-established treatment for patients with either conductive hearing loss or mixed hearing loss with only a moderate sensorineural hearing loss component [3]. The degree of the air-bone gap is of no importance since the device bypasses the ear canal and middle ear and thus the status of the pinna, ear canal and middle ear is irrelevant. However, the average BC thresholds for the frequencies 0.5, 1, 2 and 3 kHz should not exceed 60 dB and speech discriminations scores should be 60 % or better according to the manufacturer (www.entific.com).
The classic indications and contraindications are described below but owing to the characteristics of BAHA, the indications have gradually become wider which will be discussed in the end of the introductory section.

Under normal circumstances when a single-sided BAHA is fitted, the ear with the best cochlear reserve will be selected. If there is any uncertainty as to which ear is better, the patient is asked to evaluate side by applying a test rod with a BAHA behind each pinna in turns. If there is still no obvious best side, the side most convenient for the patient will be chosen; usually right-handed patients prefer to have the BAHA fitted on the right side and so forth.

The dominating indication for BAHA in adults is CSOM and in children congenital external EM.

1.4.4.1 Congenital external ear malformations

Bilateral congenital malformation of the pinna, ear canal and/or middle ear often results in bilateral maximal conductive hearing loss. At times sporadic cases occur but the presence of different malformations syndromes involving the ear, for example Treacher Collins syndrome, Pierre Robin syndrome and Goldenhar syndrome are common. Surgical reconstruction of the ear canal and middle ear defects is known to be difficult and the results are often poor with a high degree of restenosis of the ear canal and limited functional hearing in many cases. The surgical outcome in terms of social hearing has previously been studied in a series of 45 patients with external or middle EMs [57]. The results displayed a poor outcome where only 34 % obtained social hearing postoperatively and the hearing results subsided even further in the follow-up period of five years. However, with a more careful selection of surgical candidates by the use of a grading system, an assumable better hearing outcome could be expected.

To evaluate the possibilities for surgical success beforehand, a clinical ten-grade scale based on the findings in a preoperative CT scan of the temporal bone was developed by Jahrsdoerfer and associates [23]. The presence of a stapes generated 2 points, all other entrees scored 1 point each, see table 1.

Table 1. A grading system of candidacy for surgery of congenital aural atresia

Parameter	***Points***
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus/incus complex	1
Mastoid pneumatized	1
Incus-stapes connection	1
Round window	1
Appearance external ear	1
Total available points	***10***

A normal configured ear scores maximum 10 points, an isolated middle EM scores between 7 to 9 points but syndromic patients scarcely generate more than 5 to 6 points. In patients with scores of 7 points or above, there is a good chance of surgical success, i.e. postoperative speech reception thresholds of 15 to 25 dB. In patients with scores of 5 points or less reconstructive surgery might not result in enough hearing improvement. Furthermore, in order to get an indication of the severity of a middle EM, a clinical grading of the microtia can be done in the outpatient clinic where a better developed outer ear indicated a better developed middle ear [58]. CT scanning of the temporal bone can however be of value before determining potential treatment options.
If surgery will be attempted, the best ear should be operated first. Further on, surgery is usually not preformed until adolescence, however, different approaches exist at different clinics and some perform surgery earlier.

BAHA is an alternative to reconstructive ear surgery in this patient group, either on permanent basis or temporarily if reconstructive surgery is going to be carried out later.

1.4.4.2 Chronic discharging ears

The first treatment option for chronic otorrhea is revision surgery; however, multiple surgical attempts should be avoided. If hearing amplification is needed in patients with CSOM, the use of a conventional ACHA is often inappropriate and an unsafe alternative since the occlusion effect of the ear mould worsens the otorrhea problem [37]. Numerous studies [59, 60, 61, 62] have concluded that the use of BAHA significantly

reduces the episodes of discharging ear infections. In a study by Mylaneus et al. [59], the reported reduction was 94 % and in yet another study by Macnamara et al. [60] the corresponding number was 84%. The improvement in regard to ear infections seemed stable in long-term follow-up [61].
Furthermore, a persistent CSOM might result in an additional sensorineural hearing loss due to cochlear damage, especially in the high frequency region [63, 64]. Thus, in patients with CSOM resistant to medical and surgical therapy and where no successful regular hearing management is possible, BAHA fitting is considered a good option [65].

1.4.4.3 Ear canal problems

Chronic eczema or recurrent infections in the ear canal make the use of an ordinary ACHA difficult due to the ear mould. A suitable way to manage the situation is to try different materials in the ear mould or to use ventilated ear moulds. In case of continuous setbacks, a BAHA could be attempted to enable the patients to use their hearing amplification on daily basis without having the issue of itching, moisturing ear canals.

1.4.4.4 Discomfort using a conventional ACHA

Furthermore, some patients with conventional ACHA cannot tolerate the occlusion of the ear canal due to discomfort [48]. When using a BAHA instead the problem is solved, however gain might be compromised [3].

1.4.4.5 Maximal conductive hearing loss in the single hearing ear

At all times, ear surgery involves a low but certain risk of cochlear damage. In patients with a maximal conductive hearing loss on one side, for instance otosclerosis and a deaf ear on the other side, BAHA could be a good alternative to middle ear surgery.

1.4.5 Contraindications

The main contraindications for BAHA fitting are psychiatric disease, immature personality, drug and alcohol abuse and inability to follow given instructions or to participate in regular follow-up [3].

Further on, inability to maintain the implant site clean is considered a relative contraindication since good hygiene around the skin-penetrating area is essential to avoid adverse skin reactions.

Patients with severe skin disease (acne vulgaris, psoriasis, seborrhoeic eczema) or diabetes mellitus have not shown a higher incidence of implant failures or adverse skin reactions [3]. However, if severe problems with skin reaction arise, the skin around the implant site can be replaced with oral mucosa with good clinical results (Adrian Sugar, South Wales, personal communication).

Recently, the BAHA concept was tried by Sheehan et al. [66] in a group of patients with cognitive deficits i.e. Down syndrome. A high degree, 49% of soft tissue complications was encountered in this group of patients. The most common problem recorded excessive healing of the graft site with hypertrophy of soft tissue on top of the abutment. Most of the soft tissue obstacles were however rather easily resolved. In regard to osseointegration, the failure rate was acceptable and the patients and their guardians expressed high degree of satisfaction with the BAHA fitting. In conclusion, BAHA seemed like a good treatment alternative also in patients with Down syndrome when regular treatment, conventional HA or ventilation tubes has failed.

1.4.6 Surgery

1.4.6.1 Adults

Initially surgery was conducted in two stages [1]. Since the mid 90s, surgery is performed as a one stage procedure in adults as no difference has been recorded in terms of implant failures and adverse skin reactions over a five respectively eight year follow-up period [67, 68].

Adult patients are usually handled in day care units where surgery is performed under local anesthesia in the operation theatre. The most favorable position for a titanium implant, onto which a BAHA will be loaded, is considered to be 55 mm behind and 30 mm above the external ear canal [3, 34].

First, the skin over the planned operation site is prepared by shaving as well as smoothing with liquid paraffin if a dermatome is going to be used. A thin skin flap is raised, either by conventional methods or by using the dermatome and thereafter a thorough soft tissue reduction is performed. The soft tissue reduction has to be extensive to avoid later complications in terms of adverse skin reactions. A hole is drilled in the temporal line under profuse irrigation with saline solution. Generous irrigation with saline solution is of great importance to avoid heat-induced trauma that might otherwise comprise the osseointegration process. A 3.75 mm diameter threaded flange fixture of either 3 or 4 mm length is inserted into the hole. A better survival rate of the longer fixtures i.e. 4 mm has been suggested and applied whenever possible [69]. After placing the fixture, the wound is closed and the skin over the fixture site is punctured to allow the placement of an abutment onto the fixture. A healing cap is placed on top of the abutment and gauze with ointment containing corticosteroids and antibiotics is applied loosely around the abutment. The healing cap is kept in place for approximately 10 days before it is removed. After a healing period of 4 to 6 weeks the BAHA of choice can be loaded on the fixture [70].

Over the years, the surgical method has been modified somewhat to make the procedure easier and less time consuming without compromising safety issues. As already mentioned, surgery was initially performed as two-stage procedure but is now regularly done as a one-stage surgery and thus one surgical step is removed. The introduction of specially designed dermatomes, self-tapping fixtures and one-piece abutments have been time saving.

1.4.6.2 Special considerations in children

In children, special surgical considerations are to be taken into account due to the bone being more soft and immature. Further on, appositional growth of the temporal bone during adolescence might cause skin overgrowth of the abutment. Hygiene could also be an issue with cleaning difficulties around the skin-penetrating site [71].

BAHA surgery as a one-stage procedure in children has only been tried in rare cases. Due to unexpected implant failures it was soon abandoned in favor of the traditional two-stage procedure with an osseointegration period of 3 to 4 months in-between [69]. At the first stage, the skin over the implant site is incised, continuing through the subcutaneous tissue and periosteum. A 3, or preferable 4 mm fixture is placed in the temporal line, as previously described, before the soft tissue is closed. In the second procedure, the abutment is placed after a thoroughly soft tissue reduction and skin penetration. After a healing period of two to three weeks a BAHA is fitted according to clinical standards.
Since surgery is still performed as a two-stage procedure none of the new time saving methods are used in children.

In children, lack of bone might be a surgical obstacle. The insertion of a 3 mm fixture will demand at least 2.5 mm bone why a preoperative CT scan is of value in evaluation process of bone thickness and suitable placement in advance. If the bone is too thin, application of a bone generating polyetraflouroethylene (Gortex) membrane has proven to be beneficial [72]. The membrane is placed over the flange of the fixture during the first procedure and then removed at the second stage. Apart from aiding bone generation, the use of a Gortex membrane makes it possible to place the fixture more externally and thus to some degree compensate for the expected appositional growth. Before the introduction of this technique in the early 90s, lack of bone could sometimes result in that fixtures could not be installed at all. An alternative could then be a bone grafting procedure where bone from the posterior ear canal was used however this technique acquired a three-stage procedure. The bone grafting procedure can be omitted if the bone augumentation technique is used.

Yet another issue in children is the positioning of a BAHA in children with external EMs of the pinna and ear canal where reconstructive surgery might be possible. Autogenous ear reconstruction may be considered in patients with classical microtia and a relatively normal lower one-third of the ear but the decision is also dependent on patient preference and compliance [73].
A recent study conducted by Stenfelt [74] on transmission properties in cadaver heads has showed that in terms of vibration levels, the best position of a BAHA would be in the mastoid region close to the cochlea and the worst position would be at the midline of the skull. However, if reconstructive surgery of the auricle is considered the BAHA needs to be positioned further back than originally described in order not to compromise later reconstructive surgery. In a study conducted by Bajaj et al. [75], a distance of approximately 6.5-7 cm from the external auditory meatus was suggested.

Thus, in children where surgical reconstruction of the auricle is considered, it is of utmost importance to position the BAHA not too far back to compromise hearing outcome but nevertheless not to close to the ear canal to interfere with surgical outcome.

1.4.6.3 Follow-up

Regular follow-up visits in the outpatient clinic are recommended at every 3 to 6 months during the first year and then according to the patient needs. During the follow-up visits implant stability as well as status of the skin-penetrating site should be evaluated.

Most BAHA centers classify the appearance of the skin-penetrating site according to the clinical scoring system developed by Holgers et al. [76] where 0= no irritation, 1= slight redness, 2= red and moist, 3=as in 2 but granulation tissue formed, and 4 = skin irritation of such degree that the abutment has to be removed.

If adverse skin reactions appear, main treatment is intensified cleaning around the abutment with soap and water. If not sufficient, local antibiotic ointments or oral antibiotics can be prescribed and in some cases revision surgery might be needed.

The majority of percutaneous implants are stable over time. However the implant failure rate is approximately 10 % over a ten-year period where the failure rate is dependent on implant site and age at surgery [77]. The implants might be lost due to various reasons, where osseointegration failure, trauma and infections are the dominating causes. At rare occasions, the implant is removed surgically due to soft tissue reactions or chronic pains at the implant site. Development of chronic pains in the implant area is scarce and so far no satisfactory explanation has been found in why some patients develop this problem. In a study where histological examinations were done on removed implants, only a presence of inflammatory cells at varying density in the interface were noted [78].

1.4.7 New areas of use

1.4.7.1 Bilateral BAHAs

Originally, BAHA has only been prescribed for unilateral use in patients with bilateral hearing loss (BHL) otherwise fulfilling the BAHA indication criteria. Previous studies have shown that one BAHA transmits sound vibrations not only to the ipsilateral cochlea but also to the contralateral cochlea almost to the same extent why one BAHA has been considered enough for good hearing amplification [28, 29].

In recent years bilateral application has been tested in a limited number of patients and the results have been promising in patients with symmetric BC thresholds. Several studies have noted better hearing results with bilateral BAHA fitting compared to unilateral in terms of improved speech recognition in quiet and in noise, improved ability to localize sounds as well as binaural hearing to some extent [79, 80, 81, 82]. Lately, audiological outcomes with bilateral BAHAs have also been investigated in the laboratory setting by Stenfelt [83]. The results showed a theoretical benefit with better hearing thresholds from the front and better overall hearing ability from the surroundings. Moreover, bilateral fitting seemed to facilitate extraction of binaural cues and should accordingly result in improved sound localization ability and improved hearing in noise. However, due to cross hearing with bilateral BAHAs the anticipated effects would be smaller than for ACHAs.

Apart from the measured audiological benefits with bilateral BAHA fitting in comparison to unilateral, patients have also reported improved satisfaction with bilateral use. For instance, a questionnaire study from the Birmingham group found enhanced patient well-being and improved quality of life as well as a high degree of satisfaction with two aids in respect of speech perception in quiet, speech recognition in noise and localization of sounds [84].

In a recently published consensus statement on the BAHA system, bilateral BAHA fitting is advocated in young children with severe congenital conductive hearing loss and it can also be considered in adult patients after a thorough counseling and testing with a second BAHA applied on a headband [37]. However only a limited number of adults and children has so far been fitted bilaterally.

1.4.7.2 Single-sided deafness

Unilateral BAHA fitting in patients with single-sided deafness is the latest approved BAHA indication. Patients with unilateral deafness often report numerous hearing difficulties such as problems with speech perception on the deaf side, inability to localize sounds and also to understand speech in background noise. A major contributor to the perceived hearing problems is the existence of the head shadow effect which is most pronounced in the high frequency region.

Previously single-sided deafness has been considered a minor handicap and has not been rehabilitated with HAs on regular basis. However, patients complaining of more pronounced problems have been offered a conventional CROS (contralateral routing of signal) but without satisfactory results. With increasing demands from this particular group of patients unilateral BAHA has been tried in numerous patients with mainly acquired single-sided deafness.

The original theory was that fitting of a BAHA positioned close to the deaf ear would work as a direct transcranial CROS. By transmitting sounds from the deaf side to the functional cochlea the negative effects of the head shadow would supposedly be diminished and thus better hearing should be achieved in terms of expanded sound field, improved sound localization and improved speech recognition in noise [85]. Vaneecloo et al. first tried the new concept in 2 patients [86] and then later in 29 patients [87] with single-sided deafness and contralateral normal hearing. The findings were promising where BAHA fitting resulted in improvement of the previous stipulated audiological effects as well as a high level (88%) of satisfaction from a functional point of view.

Several studies have since then been performed where conventional CROS has been compared to unilateral BAHA. The effects have been investigated with both audiological methods and self-assessment questionnaires, mainly the abbreviated profile of hearing aid benefit (APHAB) [88] developed by Cox [85, 89, 90, 91, 92, 93]. Both CROS system overcame some of the negative head shadow effects and thus resulted in better speech intelligibility in noise. No improvement of sound localization ability was recorded [94]; most likely two functioning cochleae are needed for stereo effects.

Furthermore the responses from questionnaires reflected supplementary benefits with the BAHA CROS compared to the conventional CROS.

In yet another study, a group of patients with unilateral deafness and contralateral normal hearing was compared both audiologically and with questionnaires to a group of patients with single-sided deafness and contralateral moderate sensorineural hearing loss. Both the audiological results and the self-assessments were similar to previous studies; however, the benefits with BAHA CROS were more pronounced in the group of patients with moderate hearing loss in their "good ear" [95].

If BAHA implantation is considered in a patient with single-sided deafness, a trial should be arranged with a BAHA device on a softband or a steel headband, placed on the mastoid of the deaf ear [37]. How long this trial period ought to be has been debated lately and the general idea is 3-4 weeks but no consensus has been reached (IX Biomaterial Club Meeting, Val Gardena, Italy 2006).

1.4.7.3 Unilateral conductive hearing loss

Lately unilateral BAHA has not only been tried in patients with single-sided deafness but also in patients with unilateral conductive hearing loss.
The purpose with BAHA fitting in these patients has been to restore binaural hearing and thus improve their ability to localize sounds and achieve better speech recognition in noise. So far, the results have shown improvement of speech recognition in noise as well as improved sound localization ability and furthermore the patients have reported obvious benefits with BAHA in questionnaires [96, 97, 98]. The findings were most beneficial with BAHA fitting in patients with acquired conductive hearing loss but seemed more ambiguous in patients with congenital conductive hearing loss [97].

Furthermore, BAHA has also been evaluated in a group of patients with otosclerosis who could or would not undergo stapedectomy and had experienced difficulties with conventional HAs. The results were positive with improvements related mainly to comfort and cosmetics but to lesser extent to additional audiological benefits in comparison with their previous aid [99].

It may be speculated that the worse outcome in patients with congenital conductive hearing loss might be due to an imbalance in the auditory system caused by limited stimulation. In a study by Webster [100] it was shown that a unilateral conductive hearing loss resulted in an imbalance considering the size of the neurons in the two inferior colliculus where the neurons on the contralateral side were smaller than on the ipsilateral side.

Thus, in patients with unilateral conductive hearing loss unilateral BAHA makes yet an alternative. The first option to reestablish binaural hearing could still be ear surgery if possible and the second choice involves conventional HA. If none of the mentioned treatment alternatives are suitable, a BAHA might be considered after a trial period of at least two weeks with BAHA on a steel headband [97].

1.5 EAR MALFORMATIONS

EM, often as a part of a syndrome, is the prevailing BAHA indication in children why a short summary of the different syndromes associated with EMs is presented. In the study material other syndromes were present as well, but since they are not generally associated with EMs no further elaboration on those are conducted.

A malformation of the external ear i.e. the pinna, ear canal and/or middle ear is one of the consequences of a first and second branchial arch syndrome, which also involves malformations of the zygoma, orbita, lower eyelid, maxilla and mandible. The incidence of these syndromes is reported to be in the vicinity of 1 in 5 000 to 1 in 20 000 live births. The etiology is heterogeneous where sporadic mutations dominate but the syndromes can also be inherited or caused by teratogenical agents, for instance thalidomide or isotretinoin [101]. The syndromes are either unilateral (hemifacial microsomia, Goldenhar syndrome) or bilateral (Treacher Collins syndrome, Pierre Robin syndrome, Branchio-Oto-Renal syndrome (BOR)).

1.5.1 Hemifacial microsomia

Hemifacial microsomia is the most common craniofacial syndrome with a calculated incidence of 1/5600 live births [102]. The etiology of hemifacial microsomia is heterogeneous where both endogenous and exogenous factors might be responsible. A wide variety of malformations are associated with the syndrome i.e. ocular, auricular, mandibular, facial nerve and soft tissue abnormalities.

The majority of patients suffer from conductive hearing loss but sensorineural hearing loss may also be present. In a retrospective study conducted by Cravalho et al. [102] of 99 pediatric patients, the prevalence of sensorineural hearing loss was 11.1% whereas 74 % had conductive hearing loss. Furthermore the same study reported that even though the syndrome is unilateral in most cases, bilateral involvement is rather common where approximately 30 % of the pediatric patients had bilateral manifestations.

1.5.2 Goldenhar syndrome

Goldenhar syndrome or oculo-auriculo-vertebral dysplasia presents with a wide variety of clinical manifestations. The occurrence is probably mostly sporadic even though hereditary etiology might be possible [103]. The typical clinical features involve hemifacial microsomia as previously described in association with benign eye anomalies such as epibulbar dermoids or lipodermoids as well as vertebral anomalies.

Furthermore, cardiovascular malformations, clefts, teeth anomalies, mental retardation and lymphomas in corpus callosum may also accompany the syndrome [103].
Also this syndrome results in EMs and hearing impairment. Mainly the external and middle ear are engaged but CT verified malformations of the inner ear have been reported in approximately 1/3 of the cases in a study by Bisdas [104].

1.5.3 Treacher Collins syndrome

Treacher Collins syndrome or manibulofacial dystosis is caused by a mutation of chromosome 5 with an incidence of 1 per 50 000 live births. Approximately 60 % of

the cases are caused by sporadic mutations in the affected gene and the remaining 40 % is inherited in an autosomal trait with a varying penetrance [105].
The patients present with typical facial appearance characterized by antimongoloid palpebral fissures, coloboma of the lower eyelids, sunken cheekbones, preauricular fistulas, deformed pinnas and ear canal atresia, atypical hair growth extending toward the cheeks, receding chin and a large mouth. The syndrome is often associated with facial clefts, dental malocclusion and impaired hearing due to the EM.

1.5.4 Pierre Robin syndrome

The Pierre Robin syndrome consists of a triad, which includes micrognathia-retrognathia, glossoptosis and cleft palate. The etiology is heterogeneous and at least 27 different genes might be involved [106].
The syndrome might be associated with hearing impairment of conductive type where middle ear effusion is very common [107]. A light microscopy study of temporal bones from children with Pierre Robins syndrome revealed signs of middle ear infection in 100 % but a high presence of malformations involving the entire ear was also found [106]. Thus hearing impairment in this patient category might not only be caused by secretory otitis media but also by different degrees of EMs.

1.5.5 Branchio-oto-renal (BOR) syndrome

The BOR syndrome is a hereditary syndrome involving a mutation in chromosome 8 and the prevalence is approximately 1:40 000 [108]. BOR follows an autosomal dominant mode of inheritance with variable clinical expression. The syndrome is characterized by anomalies of the external, middle and/or inner ear in conjunction with preauricular sinuses, branchial cleft anomalies and varying degrees of renal dysplasia. Hearing loss is reported in 75% of the cases where 30 % have conductive hearing loss, 20 % sensorineural hearing loss and the remaining 50 % have mixed hearing loss [108].

2 AIMS

The aims of the study endeavors to obtain further knowledge of different aspects of BAHA application in children.
BAHA is since long a well-established treatment in both adults and children. Considering adults, long-term results are well documented and the indications are progressively becoming broader. Less is known of the special bone features in children that might affect surgical results and studies of new applications are lacking.
Thus, studies regarding BAHA fittings and long-term results as well as new applications in children are highly essential.

The present study was not conducted in order to give a general view of hearing habilitation/rehabilitation in patients with conductive hearing loss but merely to cover technical rehabilitation with BAHA.

The principal aims of the present study were:

To do a long-term follow-up of all children, < 16 years of age fitted with BAHA at Sahlgrenska University Hospital between 1978-1999

To do an audiological evaluation of adults fitted with bilateral BAHAs, if beneficial, it might also tried in a pediatric population

To describe the objective and subjective outcomes of different BAHA applications in children with uni- or bilateral conductive hearing loss

To investigate auditory function and self-assessed hearing problems in individuals, both adults and children with congenital single-sided external ear malformation and conductive hearing loss

Hence, the study was conducted in order to describe hearing impairment, surgical interventions and outcomes in children and to some extent adults with conductive hearing loss and BAHAs in terms of performance based measures and satisfaction measures.

3 MATERIALS AND METHODS

3.1 PATIENTS

Within the study protocol a total of 127 patients and 15 controls were investigated with assessment of their medical records, audiological measurements and/or written questionnaires. Five patients from Paper 1 were audiological tested with either uni- or bilateral BAHAs in Paper II or III.

Paper I included 41 children who were fitted with BAHA at the age of 1-16 years at Sahlgrenska University Hospital between 1978-1999. All children who had undergone surgery in order to obtain BAHA in the mentioned time frame were included in this retrospective study. The patient material consisted of 25 boys and 16 girls, where the majority was fitted with unilateral BAHA due to bilateral congenital EMs.
All adults fitted with bilateral BAHAs at Sahlgrenska University Hospital were included in a prospective study i.e. Paper II. The group consisted of 12 patients, of which 9 were females and 3 males. The main indication for bilateral BAHA application in this adult group was CSOM.
Paper III was a prospective study where the included subjects consisted of 37 children aged 6-18 years, 19 girls and 18 boys. 22 of the children were patients with either uni- or bilateral conductive hearing loss of which 16 were fitted with hearing amplification. In the majority, n=21 the etiology of their hearing impairment was a congenital EM. The remaining 15 children were included as a control group in order to validate some of the audiological methods used in the paper. The controls were all healthy and otologically normal children. All included patients were required to have age appropriate school attendance and proficiency in Swedish in extension to their hearing impairment.
Finally, Paper IV was a prospective cross-sectional study containing 57 subjects, 43 males and 14 females with single-sided malformation of the pinna, ear canal and/or middle ear causing a maximal or near maximal conductive hearing loss on the affected side.

All patients were recruited from registers at the Ear-, Nose and Throat clinics, the Audiological Units and the Craniofacial Prosthesis laboratories at Sahlgrenska University Hospital and Karolinska University Hospital as well as from the Reconstructive Plastic Surgery Department at Karolinska University Hospital. Some of the controls were recruited from the Ear, Nose and Throat Clinic at Sahlgrenska University Hospital where they had sought medical advice for benign diseases not involving ear and hearing. The remaining controls were otologically healthy students at the Alvik School.

3.2 METHODS

The study methods included mainly audiological measurements and questionnaires, which will be more closely described in the following section. However, Paper I included a throughout examination of the included children's medical records concerning their hearing impairment and BAHA fitting. Age at surgery, age at follow-up, gender, type of malformation and syndromic belonging were investigated. Indications for insertion of osseointegrated implants, number of inserted implants, number of surgical procedures and specific surgical problems at the implantation site were recorded. In 29 patients bone thickness was measured during the surgical procedure with a millimeter graded dental instrument.

3.2.1 Audiometry

All audiometric measurements were carried out in soundproof booths/rooms by an audiologist or a physician. The different stimuli were presented either by sound presentation through headphones, bone conductors and/ or loudspeakers.

In Paper II, a special test set-up was designed in collaboration with the Department of Signals and Systems at the Chalmers University of Technology. The examined person was accommodated in a sound-insulated room, with an arrangement of 12 loudspeakers spaced at 30˚ interval, thus forming a full circle, at 1 meter radius from the patient at a height equivalent to the head of a sitting person. The design was a further development of a test set-up previously used by the Nijmegen group [80, 81], with the alteration that in our study we used a full circle not only a frontal semi-circle.

In Paper III, the test set-up was modified with the intention of being more fit for pediatric use. A frontal semicircle including 5 loudspeakers spaced at 45˚ at 1 meter radius from the child was used. The test persons in both Paper II and III were instructed to hold their heads in a fixed position facing the frontal loudspeaker at each assignment. The measurements in Paper II and III involved tests with different hearing options. In Paper II, the patients were tested with unilateral BAHA on their subjective best side, the opposite side i.e. the shadow side and with bilateral BAHAs. In Paper III, the children were tested without aid or with hearing amplification uni- and bilaterally.

The patients with HAs were fitted appropriate to their hearing loss. All fittings were performed with standard methods for gain. Prior to testing the HAs were checked for proper function in both Paper II and III. The patients in Paper II were fitted with two calibrated BAHAs with equal characteristics verified by common frequency response measurement technique in practice. The volume controls were preset to give maximal amplification without causing distortion during the test. In Paper III, the children themselves set a suitable amplification level used throughout the test procedures without alterations.

3.2.1.1 Pure tone audiogram

Pure tone audiogram was measured according to clinical standards in Paper II, III and IV to obtain a current hearing status. AC thresholds were measured for the frequency range 0.25-8 kHz while the corresponding frequency range for BC thresholds were 0.5-4 kHz with some minor differences between the papers. For measurements of AC thresholds, “TDH-39 earphones” were used and for BC thresholds, a “Radioear B-71” bone transducer. The measured tone thresholds were presented as pure tone averages for the frequencies 0.5, 1 and 2 kHz (PTA (M3)) in Paper II and for the frequencies 0.5, 1, 2 and 4 kHz (PTA (M4)) in Paper II, III and IV.

3.2.1.2 *Tone thresholds in sound field*

Tone thresholds in a sound field were tested with the use of warble tones ranging from 0.5 to 8 kHz (Paper II) and from 0.5 to 4 kHz (Paper III). In Paper II, definite thresholds were tested at four different directions: at the front (0˚), at the right and left side (±90˚), and at the behind (180˚). In Paper III, thresholds were only measured at the frontal speaker position. Furthermore, sound field thresholds at 20 dB HL were considered as clinical normal and no further testing was performed if thresholds at that level were obtained at the tested frequencies.

3.2.1.3 *Speech audiometry*

Two different speech reception/recognition tests were employed in the study i.e. speech in quiet (Paper II and IV) and speech in noise (Paper II, III and IV). The speech material used throughout the study consisted of either phonemically balanced (PB) Swedish three-word sentences (Paper II and III) or Swedish PB word lists [109] (Paper IV). The used sentence material was originally developed by Hagerman [110] and consisted of ten lists including ten five-word sentences each. However, in the material used in the study the first two words of each sentence were removed.

The speech material was prerecorded on a CD and presented by a female voice for the sentence material and by a male voice for the word lists. All subjects were tested with either presentation of 50-word list or 20 three-word sentences with the task to correctly recognize and repeat the presented material.

Table II. A close display of the speech audiometry protocols used in the Paper II, III and IV.

	Paper II	**Paper III**	**Paper IV**
Sound presentation	Loudspeakers	loudspeakers	headphones
Speech material	3-word sentences	3-word sentences	one syllable word lists
Speech presentation level	most comfortable level (65-80 dB HL)	60 dB SPL	most comfortable level
S/N	adaptive method, thresholds	fixed S/N ratio, 0, +4 and +6 dB	fixed S/N ratio, + 4 dB
Aim	50 % correct	highest possible score	highest possible score

In Paper II , the speech material was presented at the frontal speaker with the aim of 50 % correct score. When testing speech reception thresholds in noise, speech weighted noise was presented at the right or left speaker (±90˚) or as surrounding noise from the remaining eleven loudspeakers.

In Paper III only speech in noise was tested with the same set of three-word sentences. The speech material was preset at 60 dB SPL while noise was presented at three different levels i.e. signal-to-noise (S/N) ratio 0, +4 and +6 dB with the aim to obtain the highest possible score.

Finally, speech in quiet was measured in Paper IV with the Swedish PB word lists previously described. In the noisy test situation, Swedish phonemically balanced words in noise (SPBN) were used where the speech-weighted noise is premixed at a fixed S/N ratio of 4 dB [111]. The presentations, both in quiet and in noise were presented through headphones at the subjects´ most comfortable level and with appropriate masking. The outcomes were subsequently compared to predicted values for each individual using the speech intelligibility index (SII) calculation described by ANSI [112]. The calculated SII scores were converted to predicted scores by the transfer function developed by Magnusson [113]. A difference between the measured and predicted score outside the 95 % CI interval determined by Hagerman [114] was considered significant.

3.2.1.4 Sound localization

Sound localization tests were performed in the horizontal plane in Paper II and III. The ability to localize sounds is known to depend on two different cues i.e. interaural difference in time (ITD) and interaural difference in sound level (ILD) where the later arises from the head shadow effect in high frequency sound above 1 kHz [115]. To test the separate influence of the two cues, a stimuli consisting of a narrow-band noise (1/3 octave) centered either below (0.5 kHz) or above I kHz (Paper II, 2 kHz and Paper III, 3 kHz), was presented three times from each speaker for a duration of one second in a random order. The subjects' task was to correctly identify the source emanating the stimuli. In Paper II the stimuli level was set at 65 dB HL and in Paper III at 50 and 60 dB SPL.

3.2.1.5 Binaural masking level difference, BMLD

BMLD is a specially designed test of binaural hearing and it was employed in Paper II. The test was carried out with bilateral BAHAs to test whether binaural hearing was achieved with bilateral BAHAs.
Basically, BMLD is yet another test of ITD and it measures a subject's ability to detect a low-frequency tone in noise. The test was originally derived for AC testing but has prior also been used in BC testing [81].
Three different conditions were tested: (1) the test tones and noise were presented equally at both ears, $S_0N_{0,}$ (2) the test tones were shifted 180˚ in phase at one ear but the noise was in phase, $S_{\pi}N_0$ and (3) the tones were in phase and the noise at both sides was 180˚ out of phase, S_0N_{π}. The test was performed with pure tones (0.25, 0.5 and 1.0 kHz) presented at 65 dB HL and a narrow band noise centered on the corresponding signal frequencies.
Thresholds of the test tone in noise were determined for all described conditions and tested frequencies where the thresholds of $S_{\pi}N_0$ and S_0N_{π} were compared to $S_0N_{0,}$ which was denoted baseline. Differences between the threshold values are referenced as "release from masking" or BMLD values.

3.2.2 Questionnaires

Paper I, III and IV included different sets of questionnaires in order to obtain the participating patients' subjective view of different aspects of BAHA fitting.

3.2.2.1 Study specific questionnaire

A study specific questionnaire was constructed exclusively for Paper I. The survey consisted of 25 questions covering the included children's opinion and usage of BAHAs. Some questions concerned basic information about the implant and the abutment, BAHA models used, previous HAs, current use of additional technical aids, maintenance of the device and contacts with the clinic and service availability. Other questions were addressing BAHA function in various listening situations and one question enquired the subject' opinion regarding the BAHA in a general context. The full questionnaire is displayed in the appendix following Paper I.

3.2.2.2 The International Outcome Inventory for Hearing Aids, IOI-HA

In Paper III, the Swedish version [116] of the International Outcome Inventory for Hearing Aids (IOI-HA) was used in the children supplied with HAs. The questionnaire is a validated seven-item survey, translated to more than 40 languages and is frequently used to measure HA outcome.
The questions target seven different domains: daily use, benefit, residual activity limitations, satisfaction, residual participation restrictions, impact on others and quality of life. Each question has five different response alternatives proceeding from worst to best outcome. In patients with mild to moderate hearing loss, scores below three indicates failure and the corresponding failure level in patients with moderate to severe hearing impairment is slightly lower [117].

3.2.2.3 Meaningful auditory integration scale and meaningful use of speech scale, MAIS & MUSS

The validated questionnaire, meaningful auditory integration scale and meaningful use of speech scale, MAIS & MUSS was used in Paper III to evaluate the parents' and homeroom teacher's opinion of the included children's hearing and communication skills in daily life situations. The questionnaire was initially developed by Robbins [118] in order to assess auditory behavior in orally habilitated children with hearing impairment.
The questionnaire contains 21 questions addressing different aspects of hearing and communication. The survey's alternatives ranged from 0 to 4 where each rating and the corresponding alternatives were never (0), rarely (1), occasionally (2), frequently (3) and always (4). The responds were thematically grouped into five different categories: hearing aid use, reaction to sounds, sound discrimination, verbal communication and speech intelligibility.
MAIS & MUSS was originally developed to be used in an interview setting but has also been used as a written questionnaire with two separate sources [119].

3.2.2.4 H-70

All patients in Paper IV completed the H-70 questionnaire at the time of the audiometry testing. The questionnaire was initially developed as a screening tool for assessing hearing problems in unselected elderly population [19]. H-70 has previously been used in numerous epidemiological studies and parts of it have been validated. The survey contains 15 questions addressing hearing problems in various communication settings as well as HA use.

3.2.3 Statistical methods

In Paper I, Fischer's exact test was executed to compare categorical data i.e. the proportion of lost implants (small sample) was compared to fixture length.

In Paper II, the obtained results were not subjected to any statistical analysis except for the speech in quiet where a paired t-test was completed to compare the outcomes of unilateral to bilateral BAHA fitting.

The statistical analyses in Paper III contained Kruskal-Wallis ANOVA, Friedman ANOVA, Sign Test and Wilcoxon Matched Pairs test. Kruskal-Wallis ANOVA was used in the analysis of the results between the included groups, which contained uneven sample sizes and were not normally distributed. When comparing data within groups, Wilcoxon Matched Pairs Test, Friedman ANOVA or Sign Test were used.

Finally in Paper IV, the statistical analyzes were executed with the following methods: Clopper Pearson, linear regression, Fischer's exact test and Kruskal-Wallis ANOVA. The Clopper Pearson method was used to calculate a 95 % confidence interval for patients with a defined interaural difference of PTAs (M4). The remaining methods in this paper were used to investigate whether the derived results from the speech recognition tests were age dependent. Linear regression was used to investigate relations between age and speech recognition scores in all included subjects and Kruskal-Wallis ANOVA and Fischer's exact test were used in comparing speech recognition outcomes in the defined age groups.

4 RESULTS

4.1 PAPER I

Long-term results and self-assessed HA usage and function in children fitted with BAHA.

Altogether 41 children were fitted with BAHA at Sahlgrenska University Hospital between 1978-1999. The recorded mean age (±SD) at the time of the first stage surgery was 8.4 ± 4.6 years and the average follow-up time 8.0 ± 5.8 years. The mean age (±SD) at the time of the data collection and completion of the questionnaire was 14.8 ± 8.3 years. However, age at the time of study varied considerable within the group where the youngest patient was 1-year-old and the oldest 37-years-old. The majority of children had bilateral malformations of the external ears and was fitted with unilateral BAHA.

4.1.1 Surgical results

The surgical intervention was performed as a two-stage procedure under general anaesthesia in the majority of cases. In three children the procedure was done in one-stage but due to implant failure in the last case of this series no more one-stage procedures have been attempted.
Surgery was without major complications in all cases. However a high proportion of contact with either the dura mater or the sigmoid sinus (70.5 %) was recorded.
The bone thickness was measured peroperatively in 29 of the children and was found to be on average 2.5 ± 0.8 mm with a tendency towards thinner bone in younger ages. Eight children were subject to bone augumentation with the Gortex membrane technique introduced in 1995 [72] due to limited amount of bone (<2.5 mm) at the first-stage.

Of 44 inserted fixtures, 20 were 4 mm long, 20 were 3 mm and in 4 cases the length was not recorded. During the follow-up period, four fixtures (9.1%) were lost; they were all 3 mm long and were lost within the first two years after implantation.

Implant stability and skin penetration site were controlled and recorded in accordance with the scoring system developed by Holgers et al. [76] in 539 visits to the outpatient clinic during the follow-up period. 92.4 % of the visits were completely reaction-free. In the remaining, 7.8 % different degrees of adverse reactions around the skin penetration site were noticed. Revision surgery was undertaken in 17.1 % of the children.

4.1.2 Questionnaire

31 of the 41 children were included in the questionnaire part of the study. The remaining children did not speak Swedish or lived at unknown addresses why they were excluded. Of the included patients, 27 responded to the questionnaire, which corresponds to a response rate of 87 %. Among the responders, 19 patients still used their BAHA and they all completed the full questionnaire. In the remaining 8 patients, the only information obtained was that they did not use BAHA at the time of the survey

and the reason why. The reason for not using BAHA was related to implant or abutment failure in four cases, one patient had received the fixture but not yet the BAHA, one patient had successful reconstructive ear surgery done, one had previous problems with recurrent ear canal infections which now had healed and finally on patient complained of acoustic feedback. Thus the actual BAHA failure rate was 18.5 % (n=5) in the questionnaire-assessed patients.

Based on the responses of the 19 active BAHA users there was a high usage rate of their BAHA and on the whole they were content with the device. Most problems in terms of BAHA function in different listening situations were reported in noisy surroundings or while talking on the phone.

4.2 PAPER II

Audiological outcomes of bilateral BAHAs in adults.

Altogether 12 adult patients with bilateral BAHAs were audiologically evaluated according to the previously described test protocol.

4.2.1 Tone thresholds

The majority (n=10) of the included patients presented with symmetrical BC PTA (M4) and the remaining two patients were slightly asymmetric.
The results of tone thresholds in the sound field showed an average improvement of 2 to 7 dB with bilateral BAHA fitting compared to unilateral fitting when warble tones were presented at the front, at the best side or at the behind. The average improvement with stimuli at the shadow side was greater i.e. 5 to 15 dB.

4.2.2 Speech audiometry

Bilateral BAHA fitting resulted in a significant improvement of 5.4 dB in speech in quiet. The speech in noise test showed an improvement where the S/N threshold with bilateral BAHAs was approximately 3 dB lower when noise was presented at the subjective best ear or as surrounding noise. However, a deterioration of 1 dB was recorded when noise was presented at the shadow side.

4.2.3 Sound localization

With unilateral BAHA the sound localization ability was close to chance level but with bilateral BAHAs there was a definite trend towards improved sound localization. In the test condition with single-sided BAHA most stimuli were perceived to origin from the aided side or slightly from behind, with bilateral BAHAs the patients could to a higher extent determine the emanating sound source.

4.2.4 BMLD

The results from the binaural test showed a tendency of achieving binaural hearing with bilateral BAHAs. However a considerable interindividual variation of the outcomes was noticed.

4.3 PAPER III

Outcomes of BAHA fitting in children and adolescents with uni- or bilateral conductive hearing loss.

In Paper III, a total of 37 children, 15 controls and 22 children with hearing impairment, were audiological tested as well as evaluated by their parents and homeroom teachers in terms of hearing and communication skills in every day life. Furthermore, the children fitted with hearing devices completed a self-assessment questionnaire in order to estimate the outcome of the HA fitting.

4.3.1 Tone thresholds

All controls displayed normal tone thresholds confirmed by normal PTAs (M4) and average sound field thresholds. A maximal or close to maximal conductive hearing impairment was noted in the children with UHL. In the sound field, the entire group with conductive UHL had normal average thresholds regardless of HA use.
In accordance with the UHL groups, both groups of children with BHL had maximal or near maximal conductive hearing loss though bilaterally. Without BAHAs, their average sound field thresholds were corresponding to a moderate to severe hearing loss. With BAHA amplification, either uni- or bilaterally their thresholds improved but were still subnormal compared to the set norm.

4.3.2 Speech audiometry

Speech recognition was tested in noise at fixed S/N ratios i.e. 0, +4, +6 dB and a set speech presentation level at 60 dB SPL. Both speech and noise were presented at the frontal speaker with the aim of highest possible correct identification of words.
Both controls and the two groups of children with UHL reached speech recognition scores close to 100 % and no difference was observed between the groups. A difference was however noted in the children fitted with HA at S/N ratio 0 dB where a significant improvement was found when they were using their regular hearing device.
The children with BHL all lacked open speech recognition without their BAHA amplification in the present test set-up. Speech recognition scores in noise were otherwise similar for the two groups of children with BHL with the use of one respectively two BAHAs. However, in the group of children fitted with two BAHAs a trend was noticed where better speech performance was seen with their usual bilateral BAHA fitting.

4.3.3 Sound localization

Sound localization ability was presented either as the ability to correct identify the emanating sound source (sound localization) or the ability to recognize the side of the emanating sound source i.e. differentiate between right and left side stimuli (sound lateralization).
All controls scored close to 100 % in regards of both sound localization and sound lateralization irrespective of presentation level, 50 or 60 dB SPL or tested frequency region, 0.5 or 3 kHz. The children with UHL displayed diminished sound localization ability in comparison to controls. Though, their results were well above chance level with a better outcome in the low frequency region. Furthermore, their ability to

differentiate between right and left sided stimuli was closer to normal. No improvement, rather a deterioration was noted when a HA was used under the test conditions.
In the children with BHL, the test could only be performed with either uni- or bilateral BAHAs since presentation levels were set at sub-threshold levels. With one BAHA they scored close to chance level, the use of bilateral BAHAs showed a trend towards better sound localization. Sound lateralization with bilateral BAHA use was even better and in close region to the results of the unilaterally impaired children.

4.3.4 Parents' and teacher's assessment, MAIS & MUSS

The response rate to this questionnaire was 80 %. The survey identified two problems areas in the hearing impaired children, both UHL and BH i.e. in reaction to sounds and with speech intelligibility. Further, the children with UHL used their HAs mainly in the scholar environment with a reported use from rarely to frequently whereas the children with BHL were reported to always use their BAHAs uni-or bilaterally.
The response pattern from the two separate sources was in good congruence.

4.3.5 Self-assessment, IOI-HA

87% of the children fitted with HAs gave the responses to the questionnaire. The three different groups of children with hearing devices scored in average above 3 in the survey. However, one exception was noted in the single item concerning impact on others, where the children with BHL fitted bilateral BAHAs scored in mean 2.5. On an individual basis, 75 % of the children with HAs scored well above 3, i.e. 4 or 5 in the separate items. Thus, the self-assessment questionnaire suggested definite treatment success with HA fitting in all groups.

4.4 PAPER IV

Auditory function and self-assessed hearing capacity in patients with congenital single-sided external EM and maximal conductive hearing loss.

The included subjects, n=57 were divided in six different age groups; 3-10 years (n=8), 11-20 years (n=17), 21-30 years (n=10), 31-40 years (n=9), 41-60 years (10) and 61-80 years (n=3). All included subjects were primarily treated for aesthetical reasons and not for their associated hearing loss. Two patients had undergone unsuccessful reconstructive ear surgery prior to the investigation. Furthermore, four patients were fitted with BAHA at the time of testing but in no case HA use exceeded 2 years.

4.4.1 Tone thresholds

The vast majority of subjects displayed air-bone gaps greater than 50 dB on the malformed side, which were considered equivalent to a single-sided maximal conductive hearing loss.
A comparison between each subject's normal ear AC PTA (M4) and the malformed ear BC PTA (M4) was carried out to access hearing threshold function. No difference was found in the majority of cases but in 16 % worse thresholds were found on the malformed side. In the group with worse outcome on the malformed side, a high rate of unilateral craniofacial malformation syndromes was noted.

Furthermore, a possible protective effect of conductive UHL was investigated in regards of noise induced hearing loss but no such effect could be seen within the study.

4.4.2 Speech audiometry

The outcomes from the speech recognition test in quiet and noise were compared to predicted values calculated for each individual. A good congruence between the measured and predicted scores was seen on the normal side. However, the results from the malformed side showed a discrepancy between the measured and predicted score in over 50 % of the cases with worse outcome on the malformed side than predicted.

4.4.3 Self-assessment questionnaire, H-70

The subjects reported a high degree of self-assessed hearing problems. In general, 77 % of the included subjects perceived impaired hearing capacity. Some of the reported problem areas within the survey were in conversation in noisy surroundings as well as in sound localization ability. Furthermore, a high prevalence, 54% of tinnitus within the group was reported.

5 DISCUSSION

5.1 PATIENTS

The present study focused on different aspects of BAHA application in children why mainly patients < 18 years of age i.e. 102 children were included. However, adults were included in Paper II and IV. In Paper II, audiological outcomes were measured in adults fitted with bilateral BAHAs for various reasons prior to the investigation. Since bilateral BAHA fitting is a new area of BAHA use, a study performed in adults ought to precede a similar study conducted in children. Furthermore, little is known of the consequences in the auditory system of a congenital external EM with associated maximal conductive hearing loss causing limited stimulation in the affected side. In Paper IV this issue was addressed why individuals with a wide age range, 3-80 years were included in order to observe hearing function at different ages.
All participating BAHA patients were prior to the study fitted with either BAHA uni-or bilaterally, thus no surgical treatment or other interventions were included within the study protocol.

A male dominance in the material was observed, where 86 subjects were male and 56 female corresponding to a male /female ratio of 1.5 to 1. As the study covered investigations concerning BAHA mainly in children as well as auditory function in patients, both adults and children with single-sided external EMs the observed gender distribution is thought to be representative since congenital malformations are known to be more frequent in males with a ratio of approximately 2 to 1 [120].

The study patients were recruited from registers at the Ear-, Nose and Throat clinics, the Audiological Units and the Craniofacial Prosthesis laboratories at Sahlgrenska and Karolinska University Hospital as well as from the Reconstructive Plastic Surgery Department at Karolinska University Hospital. They had all sought medical advice for either hearing loss or aesthetic reasons.
Both hospitals from where the subjects were recruited were well familiar with the BAHA concept and thus BAHA might have been offered to a higher extent than in other clinics.
Among the patients who were primarily treated for hearing related problems, a selection might also have been possible to some extent. The patients, both adults and children with BHL who were fitted with bilateral BAHAs as well as the children fitted with unilateral HA, either an ACHA or a BAHA had actively sought more extensive hearing rehabilitation than is normally offered in clinical practice. Thus, these patients may not be a representative sample of subjects with conductive hearing impairment uni- or bilaterally.
However, in the patient group who was primarily treated for aesthetic reasons, selection bias was not considered to have been an issue. All patients in this group suffered from single-sided external EM where either reconstructive surgery of the pinna or episthesis were considered, and their associated hearing problems were primarily not addressed. Thus, they should represent an unselected population with single-sided congenital craniofacial malformation and unilateral maximal conductive hearing loss.

Furthermore, in Paper IV, patients with a wide age span i.e. 3-80 years were included. The majority of this group was 40 years or younger suggesting that they seek medical treatment for aesthetical corrections to a higher extent.

The vast majority of included patients had conductive hearing loss, either uni- or bilaterally without any sensorineural component. However, a minority of the subjects displayed mixed hearing loss with an additional mild to moderate sensorineural hearing impairment.

In Paper II, audiological outcomes with bilateral BAHA fitting were compared to unilateral BAHA. Among the included patients, two patients were fitted with bilateral BAHAs despite only mild hearing loss according to their PTA (M3) with limited air-bone gaps on the second implanted sided. Under normal circumstances, patients with mild hearing loss are rarely fitted with HAs regardless of etiology but in these particular cases their hearing fluctuated due to frequent infections why hearing amplification was at times necessary.

Hearing impairment is preferably rehabilitated with ACHAs since the AC route of sounds is more effective than the BC route. BAHA is; thus, only an alternative in a small portion of patients with hearing impairment where ACHAs are not feasible or appropriate due to EMs or chronic ear infections.

The present study concerns primarily different aspect of BAHA fitting where the outcomes of new indications are studied in an adult and pediatric population. All available adults and children with BHL and bilateral BAHAs as well as all children with UHL and unilateral HA from the two major cities in Sweden were included. Due to the limited number of patients in the studies only trends can be identified without power to reach statistic significance. The problem with small patient populations is an obstacle throughout this field of research. Most BAHA centers perform similar tests on their respective patient material in order to enable comparisons of the results between the different centers and thus add more knowledge about how BAHA could be used in an optimal way.

5.2 METHODS

Several well-established audiological measurements were executed in the present study. In addition, validated questionnaires were included in Paper III and IV. However, in Paper I, a study specific questionnaire was used.

In this section some of the methods will be discussed more closely.

5.2.1 Tone thresholds

Tone thresholds in a sound field were measured in both Paper II and III, however the purpose and method differed between the papers. In Paper II, the test was part of the audiological assessments of possible benefits with bilateral BAHA fitting why actual thresholds were obtained at four different directions with unilateral BAHA on either the best or the shadow side and with bilateral BAHAs. In Paper III, however, a modified test protocol was used in order to limit the measurements in the pediatric material. In this paper, tone thresholds in the sound field were tested with the mere purpose to control the accuracy of the previously obtained pure tons thresholds as well as to act as a tool to control appropriate gain of the used HAs.

5.2.2 Speech audiometry

The present study employed two different speech reception/recognition tests i.e. speech in quiet (Paper II and IV) and in noise (Paper II, III and IV) with the use of either PB three-word sentences or word lists.
Hagerman [110] has developed a Swedish speech material containing ten five-word sentence lists. This material has been validated in adults but not in a pediatric population. To date validated Swedish speech material for pediatric use is lacking.
In Paper II and III, Hagerman's three-word sentences were used i.e. the two first words of each sentence were removed. No published validation has been presented with the three-word material; however it has previously been used in approximately 350 children with similar equality in intelligibility as the five-word sentences (personal communication, Tomas Tengstrand, master of engineering, the Audiological Unit at Sahlgrenska University Hospital, Göteborg, Sweden).
In common clinical practise Hagerman's three or five word sentences are mainly used in adults with an adaptive method in order to detect a threshold value of the S/N ratio for certain speech reception, usually 40 or 50 %. In Paper II, which included adults fitted with bilateral BAHAs, the test was used with three-word sentences and estimation of speech reception thresholds at 50 %.
In Paper III the test choice was set at Hagerman's three-word sentences at three fixed S/N ratios i.e. 0. +4 and +6 dB since no validated speech material for pediatric use are available. The results of the chosen test came close to the ceiling effect for the controls and children with UHL regardless of HA use why a more difficult speech recognition test with higher noise level might have been a better choice in order to differentiate between these groups. In children with BHL, the test seemed more appropriate as the results were below the ceiling level indicating a better choice of S/N ratio to discriminate between different hearing options i.e. with or without BAHAs.
In Sweden, speech recognition ability is usually screened with PB lists why this method was used in paper IV where the included subjects were tested by various audiologists at multiple hearing centres. In Paper II and III, all tests were completed by only two persons i.e. one physician and one audiologist.

5.2.3 Questionnaires

In Paper I, a specially designed questionnaire was constructed. However, in Paper III and IV validated questionnaires were used i.e. MAIS & MUSS, IOI-HA and H-70. The selection of the most appropriate questionnaires can always be subject to discussion.

At the time of the long-term evaluation of BAHA in children, only a limited number of studies [13, 47, 56, 121, 122] had been published where questionnaires were used in conjunction with BAHA outcome.
One questionnaire, which was previously used in a pediatric material fitted with BAHA was constructed by the Birmingham group [13]. This survey contained 15 questions addressing the child's use of BAHA and attitude towards the device. The questionnaire was used to large extent as a raw model when constructing the study specific questionnaire used in Paper I.
Since then more than 15 different validated questionnaires and several "home-made" questionnaires have been used to assess the subjective outcomes of BAHAs. The

surveys address different categories such as quality of life, health related quality of life, disability assessment, communication problems as well as specific hearing impairment. The three most frequently used questionnaires have so far been the Glasgow Benefit Inventory [123] developed by Robinsson et al. [84, 92, 124, 125, 126], the Abbreviated Profile of Hearing Aid Benefit [88] developed by Cox [85, 89 90, 91, 92, 95] and the Nijmegen questionnaire [122] developed by Mylaneus [59, 61, 127].
Thus, a wide assortment of questionnaires is available to date and no agreement exists in terms of which questionnaire to use when evaluating BAHA. In future, a consensus regarding questionnaire use in BAHA patients might be of value in order to simplify comparisons of outcomes between the different BAHA centers, patient groups, interventions and indications.
In the present study the constructed questionnaire used in paper I addressed primarily issues related to the BAHA and the hearing impairment. Furthermore, in Paper III the questionnaires assessing hearing and communication skills as well as HA outcome were chosen. Finally, in Paper IV the included questionnaire was used as a screening tool thus addressing disability assessment.

5.3 SURGERY RELATED OUTCOMES

Paper I dealt with long-term surgical outcomes of BAHA implantation in a pediatric material.

BAHA implantation in children requires some special considerations due to more immature bone and appositional growth of the temporal bone. Furthermore, the estimated survival time of an implant in a child needs to be extended in comparison to adults due to longer life expectancy. At present, no data exist on expected survival time of osseointegrated implants in children for time intervals extending eight years.
The recorded implant failure within the study was 9.1% (4 implants) during the observation period of 8.0 ± 5.8 years and all implants were lost during the first two years after surgery. The noted implant failure rate was well in accordance with previous studies on children supplied with fixtures for BAHAs or BAEs, where the failure rate was ranging from 5-20% [11, 12, 14, 15, 16, 69]. The failure rate also corresponded well to an adult material where the long-term implant failure was 5-10 % over a 10-year follow-up period [129]. The majority of implants in the mentioned studies were lost early due to failed osseointegration but some were lost later due to trauma. In no case loss was due to severe adverse skin reaction.
Observations around the skin-penetrating site in the present study were reaction-free in 92.4 % and if reactions appeared they were generally mild (grade 1). These findings were also well in accordance with earlier studies in both adults and children [15, 22]. In neither the present study nor in earlier studies, was there any case of severe adverse skin reaction to the degree that the implant had to be removed.
Thus, after osseointegration has occurred the implants seemed stable over time and the degree of adverse skin reactions acceptable.

Another issue implanting children is lack of available bone at the implantation site. In the present study, bone thickness was measured peroperatively in 29 of the children and was found to be on average 2.5 mm ± 0.8 mm with a tendency towards thinner bone in younger ages. A minimum of 2.5 mm is considered necessary to implant a 3 mm

fixture. In patients with thin bone two different techniques have been described i.e. bone transplantation where extra bone is added or bone augumentation with the use of a Gortex membrane in order to induce bone growth. Furthermore, if there is any doubt about the long-term viability of the fixture a second “sleeper fixture” is suggested [13].
In the present study, ten children presented with thinner bone than 2.5 mm. Eight of these were operated with the bone augumentation technique. The remaining patients were implanted before the introduction of Gortex membranes and thus were not subject to this technique. No implants losses were recorded either in the bone augumentated group or in the two remaining children with thinner bone than 2.5 mm.
In the group with implant failures, bone thickness was only measured in one child and was found to be 3 mm. Thus thinner bone might be a surgical obstacle, especially in younger children but the use of Gortex membranes makes implantation also possible in very young patients. In this series of patients, the youngest patient was operated successfully at the age of 18 months.
At present, implantation is however not recommended in children < 2-3 years of age but the real limit is not known [37]. A promising alternative in the younger children is BAHA fitting on softband [53] until surgery can be performed.

The problem with implant failures is most likely a complex issue where fixture length may be one factor of importance for implant success. The manufacturer recommends the placement of a 4 mm fixture whenever possible. In the present study, four implants were lost and they were all 3 mm long suggesting better survival of longer implants. This was in accordance with a previous study from our group where 100 patients were supplied with fixtures for either BAHAs or BAEs. The implant failure rate in this study was 5.8 % of 170 inserted fixtures where 90 % of the lost implants were 3 mm long [69].

Another surgical obstacle when implanting children is the appositional growth of the temporal bone, which may result in formation of subcutaneous tissue and bone around the implant. Revision surgery, where additional soft tissue and bone is reduced, is sometimes necessary due to a shorter distance between the skin surface and the abutment.
In the present study, revision surgery was performed in 17.1 % of the children. In previous studies in children, the performance of revision surgery varied between 10-25 % [12, 14, 15]. The problem seems to be most aggressive in children between 6 to 12 years of age and then it subsides [69].
In order to reduce the need for revision surgery as well as trying to diminish the failure rate, the Gothenburg center has started to implant 4 mm long fixtures in conjunction with Gortex membranes in all children regardless of bone thickness.

5.4 AUDIOMETRY

Audiometric measurements were performed in Paper II, III and IV. The chosen methods for the different tests have already been closely discussed in this section why only results are considered here.
In Paper II and III, bilateral BAHA fitting were investigated from an audiological point of view, however, due to different methodology in the studies only results from the sound localization tests can be compared.

In future studies, efforts should be made to construct and validate test material suitable for all ages. Constructing appropriate tests where ceiling effects can be avoided would be a real challenge.

5.4.1 Tone thresholds

Pure tone thresholds were measured according to clinical standards in Paper II, III and IV in order to obtain current hearing status. Previous audiograms were available for most patients in Paper II and III and for patients younger than 20 years in Paper IV. The remaining patients in Paper IV had not been audiological tested in recent time.
The patients' pure tone thresholds were symmetric in the majority of individuals in Paper II. In this group, CSOM was the dominating indication for BAHA and the hearing loss was either mixed or conductive. In Paper III the children had maximal conductive UHL or BHL and in Paper IV all included subjects had maximal or near maximal conductive UHL.

Tone thresholds were also tested in a sound field in Paper II and III. In Paper II, the test was used to determine any benefits with bilateral BAHA fitting with sound presentation at different directions in the sound field. The results showed a positive effect with bilateral BAHA fitting i.e. gain of 5-15 dB if tones were presented at the shadow side. This was most likely due to the removal of the head shadow effect. Furthermore, an improvement of approximately 2-7 dB was seen for the three other tone presentations i.e. at front, at the best side or at the behind. This effect was considered to be due to an energy doubling and a double increase of signal amplitude in phase.
In Paper III, tone thresholds measurements were merely included to confirm the result from the previous pure tone audiogram as well as to check for appropriate HA function before proceeding with the remaining test protocol.
No additional effect was found in the children with UHL when fitted with their regular HA, most often a BAHA, indicating that the threshold levels were dependent on the better ear's capacity. In the children with BHL the use of a unilateral BAHA resulted in hearing thresholds close to the set norm of 20 dB HL and no extra threshold gain was recorded with bilateral fitting.
In Paper IV, an intraindividual comparison was made between the normal ear's AC PTA (M4) and the malformed ear's BC PTA (M4). In the vast majority of subjects no significant difference was noted between the ears indicating a sufficient competence to detect pure tones if presented at adequate levels. In 16% of the patients, worse BC PTAs (M4) were noted on the malformed side. In this relatively small group a high prevalence of unilateral craniofacial malformation syndromes was noted why related inner EMs could be suspected. Bisdas et al. [104] has previously showed a high proportion (36%) of associated inner EMs in extension to external EMs in patients with Goldenhar syndrome in a CT based study.
A possible protective effect of noise induced trauma in patients with conductive UHL has previously been studied [130, 131] and the results have been ambiguous. To address this issue, a subanalysis was performed where self-reported noise exposure was correlated to the AC thresholds on the normal ear and the BC threshold at the malformed ear at 4 kHz. In this present study only a limited number of patients were subject to noise exposure and no effect, either positive or negative could be seen within this group.

Thus, to investigate any influence of a conductive UHL on noise induced hearing loss larger patient material is needed.

5.4.2 Speech audiometry

Speech reception/recognition in quiet was measured in Paper II and IV and in noise in Paper II, III and IV.

5.4.2.1 Paper II

An improvement of 5.4 dB of the speech reception thresholds was obtained with bilateral BAHAs in comparison to unilateral fitting. Similar results have previously been reported [79, 80, 81]. The observed improvement was considered as an effect of energy summation at the cochlear level as well as a dichotic summation in the central auditory pathways.
When measuring speech reception in noise an improvement was found with bilateral BAHA fitting in comparison to unilateral if noise was presented at the best side or as a surrounding noise. If noise was presented at the shadow side however, less noise was tolerated to fulfill the aim due to the removal of the head shadow effect. Thus, bilateral BAHA fitting seems beneficial if noise emanates from multiple sources or as a diffuse field, which probably correlates best to noisy surroundings in every day life. However, in the case of a well-defined noise source, the use of a unilateral BAHA on the opposite side of the noise source could be more favorable.
The results correlate well to previously conducted measurements in Nijmegen [80, 81].

5.4.2.2 Paper III

The interpretation of the results from the speech recognition test in paper III was somewhat intricate due to the described ceiling effect. All controls and children with UHL regardless of HA use in the test scenario scored close to maximum. A significant improvement was however obtained with HA use in the UHL group of children at the most difficult test situation i.e. S/N ratio 0 dB. Hence, HA fitting might be beneficial in more noisy surroundings.
In previous studies, where speech recognition was tested with separate sources for speech and noise in adults with acquired or congenital conductive UHL improvements with unilateral BAHA fitting compared to the unaided situation have been reported [97, 98].
In the children with BHL, speech recognition performance did not differ between the group that was regularly fitted with unilateral BAHA and the group fitted with bilateral BAHAs. A trend within the bilaterally fitted group was however noticed where speech recognition scores improved with bilateral fitting in comparison to unilateral BAHA. Hence, in adults benefits with bilateral BAHA fitting in regards to improved speech reception thresholds has been shown but the effect is still unclear in a pediatric population. Larger material is needed for further evaluation.

5.4.2.3 Paper IV

A comparison between measured and predicted speech recognition scores in quiet and in noise was conducted in paper IV. A majority of the subjects displayed significantly worse outcome on the malformed side than was anticipated, in both test conditions.

Hence, the poorer performance suggested disturbed hearing function though it was unclear whether this was an inner ear malfunction or a disturbance of the central auditory pathways.

5.4.3 Sound localization

The results from the sound localization test showed a distinct trend towards improved sound localization ability in both adults (Paper II) and children (Paper III) fitted with bilateral BAHAs compared to unilateral. No difference was noted at presentation of low frequency stimuli versus the high frequency stimuli; thus, the patients seemed able to use both ITD and ILD cues for localizing sounds.
Among the children with UHL no benefit was seen with HA application. Rather their sound localization/ sound lateralization ability was better without their hearing amplification. Previous studies have shown better sound localization ability in adults with acquired conductive UHL fitted with BAHA whereas the results have been more ambiguous in patients with congenital conductive UHL and single-sided BAHA fitting.

5.4.4 BMLD

The BMLD test has previously been used to test binaural hearing capacity in 25 adult patients fitted with bilateral BAHAs [81]. The release from masking in Bosman's study was greatest at the lowest tested frequency, 0.5 kHz where the BMLD effect was 6.6 dB for the $S_{\pi}N_0$ condition relative to the baseline, S_0N_0. The effect decreased with increasing frequency. Over all the results were less pronounced than those normally found in otologically normal individuals but were better than in individuals with sensorineural hearing loss [81].
In the present study there was wide individual variation, however a trend towards release from masking in the vicinity of 3-5 dB was noted. Thus the obtained results both from the present study and from the previous study by Bosman indicated a presence of binaural hearing at least to some extent with bilateral BAHA fitting in adults.

5.5 QUESTIONNAIRES

The study contained four different questionnaires in order to gather information about satisfaction measures as well as to investigate whether any improvement in activity, participation and quality in life was the outcome of HA fitting.

5.5.1 Study specific questionnaire

In Paper I, a study specific questionnaire was mailed to 31 of the retrospectively evaluated children, where either the children completed the survey themselves or with some assistance from a parent.
Eight of the 27 responders reported no usage of the BAHA at the time of the survey. The reason for not using BAHA was related to the device or the implant in five of the patients and only these were regarded as failures. The remaining three patients were not regarded as "true" BAHA failures, for instance one child had only received the implant but was not yet fitted with BAHA, one patient had undergone successful reconstructive ear surgery with social hearing and finally one child previously successfully used BAHA but the patient was currently using ACHA instead. No information was

accessible at time of the study, whether the four non-responders or the remaining ten patients lost to follow-up used their BAHAs.
Among the 19 BAHA users, the great majority expressed contentment with the device and used it on daily basis. The reported function pattern of the BAHA in different listening situations displayed mainly one problem area i.e. listening in noisy situations where close to 50 % experienced the function as either very unsatisfactory or unsatisfactory. These results were well in accordance with a study from the Birmingham group [132] on both adults and children fitted with BAHAs where the same type of questionnaire was used.
Three patients reported very unsatisfactory or unsatisfactory function with BAHA while talking on the phone and they commented that the problems occurred only with cellular phones. Problems with annoying effects due to electromagnetic interference have previously been reported with the BAHA Classic device [133] but not with BAHA Compact [134]. Among the patients complaining of problems with cellular phones in the present study two were fitted with BAHA Classic and one had a BAHA Cordelle. Thus, in the newer BAHA models this problem seems to have been solved.
The skin condition at the skin-penetrating site as well as implant stability was checked on regular basis in the out patient clinic. As previously mentioned the great majority of observations were graded as completely reaction-free. However, 68 % of the children reported at least one episode of adverse skin reaction. Papsin et al. has previously reported a corresponding incidence of 82 % in their pediatric material [12]. Thus, the occurrence of temporary adverse skin reactions is common but nevertheless the skin site was graded as completely reaction free at the majority of follow-up visits suggesting good healing capacity.

5.5.2 MAIS & MUSS

The results from parents' and teacher's assessment on the children's hearing and communication skills in daily life revealed two main problem areas, i.e. in reactions to sounds and with speech intelligibility. Most problems were reported within the theme of reaction to sounds, where no difference could be seen between the children with UHL or BHL. Concerning the speech intelligibility, a higher extent of reported problems was documented among the children with BHL, but even the children UHL scored lower than the control group.
Thus, several problems in hearing and communication skills were reported not only in the children with BHL but also in the children with UHL. The report [25] on the consequences of UHL in children supported the present findings that also children with UHL are affected by their hearing impairment. Hence, hearing intervention may be of importance also in children with UHL.

5.5.3 IOI-HA

In Paper III, the children fitted with HAs completed the written questionnaire. Most children completed the questionnaire themselves but some of the younger children received some help by their parents in explaining the questions to them.
In general both the children with UHL and BHL supplied with hearing amplification either uni-or bilaterally disclosed scores according with treatment success due to the described protocol interpretation [117].

Hence, the self-assessed HA benefit was in favour of hearing amplification as opposed to no technical hearing intervention in all three groups supplied with HAs.

5.5.4 H-70

H-70 was used as a screening tool for self-assessed hearing problems in the subjects with single-sided external EM and associated conductive hearing loss in Paper IV. The survey displayed an extensive degree of general hearing difficulties in the study population compared to a general adult population [18]. The most problematic situations reported were conversation with many persons, conversation in traffic noise and sound localization.
Thus, single-sided maximal or close to maximal conductive hearing loss result in a high degree of self-assessed hearing problems why hearing rehabilitations efforts might be considered in this cohort. To date, experience is limited on rehabilitation outcomes in this particular patient group but both reconstructive ear surgery in selected cases and unilateral BAHA fitting look promising [98, 135].

5.6 CLINICAL CONSIDERATIONS

At present, patients with BHL of conductive or mixed type suitable for the BAHA concept might be considered for bilateral BAHAs. The recent consensus [37] on the BAHA system, based on the results from Paper II and from the Nijmegen group[81], suggested bilateral BAHAs on softband temporarily in children awaiting surgical treatment with bilateral BAHAs. In adults with symmetrical cochlear function a second BAHA can be offered and tried on a headband before final decision of an additional implant. Thus, the option of bilateral BAHAs should be explored further.

In patients, both adults and children with UHL HAs might be beneficial. Due to the described consequences of UHL in children it has been advocated that these children should be supplied with a HA as soon as possible [37]. In patients with UHL of conductive type, BAHA makes an alternative to ACHA. Finally, HAs might also be offered on a trial basis to patients regardless of age with congenital conductive UHL due to reduced auditory function in conjunction with a high rate of self-assessed hearing problems.

6 CONCLUSION

This study of children and adults with uni- or bilateral conductive hearing loss has employed descriptive methods as well as performance based and satisfaction measures to attend different aspects of consequences and technical rehabilitation with BAHA.

Based on the main findings of the study the following conclusions were drawn:

- Long-term results of BAHA fitting in children displayed the same frequency of implant failures and adverse skin reactions as in adults. Furthermore, benefits and function patterns in the investigated pediatric material were also in parity with adults why BAHA was assessed as a safe and well functioning technical aid also in children.

- An audiological evaluation of bilateral BAHA fitting in comparison to unilateral in adults revealed auditory benefits in terms of improved speech reception in both quiet and in noise, enhanced sound localization ability as well as some degree of binaural hearing. Thus, bilateral BAHAs could be considered in adult patients. The paper set a sound platform for conducting a similar study in children.

- Audiological outcomes in the pediatric material showed a slight improvement of speech recognition in noise but no positive effect was found in sound localization ability in children with conductive UHL fitted with a hearing device. In children with conductive BHL, a trend with improved speech recognition in noise as well as improved sound localization was noticed when fitted with two BAHAs instead of one. Problems were recognized in the children's reactions to sound and in their speech intelligibility. All children with HA intervention reported a positive outcome with their devices. Thus unilateral HA in children with conductive UHL as well as bilateral BAHAs in children with conductive BHL seemed beneficial but further studies are needed in a larger population.

- Single-sided external EM with associated conductive hearing loss resulted in compromised auditory function in terms of supra thresholds processing on the malformed side. Furthermore, this cohort reported a high degree of self-assessed hearing difficulties why technical hearing intervention, reconstructive ear surgery and other rehabilitation support should be considered.

7 FUTURE STUDIES

In future, the topic of this thesis should be elaborated further where a few suggestions are made below.

A methodology study is essential in order to validate a Swedish speech material also for pediatric use which to date is lacking. The Hagerman three-word sentences with fixed S/N ratio seemed promising but further evaluation is needed. Various fixed S/N ratios should be tested in order to find the ultimate S/N ratio to use. Most probably the fixed S/N ratio should be set differently when testing children with mild hearing loss i.e. conductive UHL in contrast to children with moderate to severe hearing loss i.e. conductive BHL to avoid ceiling effects.

The auditory function in subjects with single-sided EM and maximal conductive hearing loss seems to be diminished. Additional studies are required to investigate the effects more closely in order to obtain further knowledge of the patophysiology.
At present, guidelines about audiological interventions are lacking in this particular group of patients and some patients are subject to intervention and some are not. A randomized multi-center study including all newly diagnosed children with single-side external EM and maximal conductive hearing loss might be an option. The included subjects would be randomized in two groups, one without technical hearing intervention and one with HA fitting, either BAHA or if possible ACHA. CT scans of the temporal bone as well as repetitive psychoacoustical measurements should be performed.

Furthermore, the effects of HA fitting, mainly BAHA should be further evaluated in patients, both adults and children with conductive UHL. The subjects previously included in paper IV could be invited to test a BAHA mounted on either a softband or a steel headband for a trial period of three months and subsequently evaluated with audiological methods as well as with self-assessment questionnaires.

Finally, a larger and preferably controlled study is needed to evaluate the audiological effects of bilateral BAHA fitting in a younger material. Since the number of children with conductive BHL suitable for the BAHA concept is scarce, a multicenter study including some of the larger BAHA centres for instance Göteborg/Stockholm, Nijmegen and Birmingham would be favourable. However, a preceding methodology study should be performed in order to find suitable reliable tests for this type of pediatric material.

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10 SVENSK SAMMANFATTNING

BENFÖRANKRAD HÖRAPPARAT (BAHA) HOS BARN

Bakgrund

Sedan introduktionen av den benförankrade hörapparaten, BAHA 1977 har mer än 25 000 patienter försetts med BAHA globalt och antalet stiger successivt. Tekniken bygger på osseointegration dvs. ett titanimplantat inkorporeras i levande ben utan att någon rörelse kan ske mellan de ingående komponenterna. Titanimplantat placeras i skallbenet bakom örat i samband med ett mindre kirurgiskt ingrepp. På implantatet kopplas en BAHA som leder ljud till de båda inneröronen via skallbenet.
Indikationer för BAHA utgörs av medfödd missbildning av hörselgång och/eller mellanöra med samtidig hörselnedsättning samt kroniskt rinnande öron som omöjliggör användandet av en konventionell hörapparat som helt eller delvis är placerad i hörselgången. Initialt förskrevs BAHA endast till vuxna men är sedan flera år även godkänt för barn. Erfarenhet angående indikation, operationsteknik och framför allt komplikationer samt långtidsresultat är stor hos vuxna men mindre hos barn.
Vanligen förskrivs BAHA för ensidigt bruk även om de flesta patienter har dubbelsidig hörselnedsättning. Vid användning av konventionell luftledd hörapparat förstärks ljudet endast på det hörapparatförsedda örat varför man normalt förskriver dubbelsidiga apparater vid dubbelsidig hörselnedsättning. Med BAHA förstärks ljudet emellertid nästan i samma grad i de båda inneröronen. Detta fenomen har lett till att man normalt har försett patienter med dubbelsidig hörselnedsättning med endast en BAHA då man ansett att detta är tillräckligt. Ett par mindre studier på vuxna samt barn visar dock att utprovning av dubbelsidig BAHA kan vara av värde.
Vidare har man tidigare ansett att en medfödd ensidig hörselnedsättning oavsett orsak inte behöver rehabiliteras då hörsel på ett öra har ansetts som tillräckligt. Men som i fallet med ensidig BAHA vid dubbelsidig hörselnedsättning har dessa individer nedsatt riktningshörsel, nedsatt eller upphävd förmåga till binauralt hörande samt har svårigheter att uppfatta tal framför allt i omgivning med störljud i bakgrunden. En nyligen publicerad metanalys visar dessutom att barn med ensidig hörselnedsättning har mer problem med skolgången än jämnåriga normal hörande samt att det föreligger en risk för försening av språk- och talutvecklingen.

Patientmaterial ingående i avhandlingen:

Patientmaterialet bestod av sammanlagt 127 patienter samt 15 kontroller där den övervägande delen utgjordes av individer under 18 år. Patienterna rekryterades ur patientregister på öron-, näs- och halsklinikerna, hörselvårdsmottagningarna samt ansiktsprotesmottagningarna vid Sahlgrenska samt Karolinska Universitets Sjukhuset och från avdelningen för rekonstruktiv plastik kirurgi vid Karolinska Universitets Sjukhuset. Samtliga inkluderade patienter hade antingen ensidig eller dubbelsidig hörselnedsättning av ledningsfelstyp. Kontrollerna bestod av öronfriska barn.

Studie I: Metod: En retrospektiv studie av alla barn (n=41) som hade försetts med BAHA på öron-, näs- och halskliniken vid Sahlgrenska Universitets Sjukhuset mellan 1978-1999. Journalhandlingar gicks igenom beträffande indikation, kirurgi och e v komplikationer samt uppföljning. Bentjockleken mättes i samband med operationen hos 29 patienter. Vidare besvarades en enkät där barnen och/eller föräldrarna fick värdera hörapparaten. Resultat: Studien visade att frekvensen av implantatförluster, hudreaktioner samt BAHAs funktionsmönster är likartad hos barn och vuxna.

Studie II : Metod: Samtliga vuxna med dubbelsidig BAHA vid Sahlgrenska Universitets Sjukhuset inkluderades i en prospektiv audiologisk utvärdering av ensidig jämfört med dubbelsidig BAHA. Hörseltester, både konventionella samt av forskningsgruppen special utvecklade, utfördes av tontrösklar, taluppfattning i tyst samt i brus, riktningshörsel samt binauralt hörande. Resultat: Dubbelsidig BAHA uppvisade bättre resultat än ensidig i form av förbättrad taluppfattning i både tyst och i brus, förbättrad riktningshörselförmåga samt till viss del även binauralt hörande. Resultaten ligger till grund för vidare studier i ett yngre material.

Studie III: Metod: En prospektiv studie inkluderande 22 barn med ensidigt eller dubbelsidigt ledningsfel som antingen var obehandlade eller försedda med en eller två BAHA samt 15 öronfriska kontroller utfördes. Audiologiska mätningar inkluderade testning av tontrösklar, taluppfattning i brus samt riktningshörsel. I enkäter kartlades problemområden inom hörsel och kommunikation samt självupplevd nytta av utprovat hörhjälpmedel. Resultat: Både ensidig och dubbelsidig hörselnedsättning resulterade i problem inom reaktioner på ljud samt vid språkförståelse. Utprovning av hörapparat, antingen BAHA eller konventionell hörapparat vid ensidigt ledningsfel ledde till förbättring av taluppfattningsförmågan i situationen med mest störljud men däremot sågs ingen förbättring av riktningshörseln. Dubbelsidig BAHA vid dubbelsidig hörselnedsättning tenderade till bättre taluppfattning i bullrig miljö samt förbättrad riktningshörsel. Samtliga barn försedda med hörapparat rapporterade positiv effekt av utprovningen.

Studie IV: Metod: Sammanlagt 57 försökspersoner i åldrarna 3-80 år med medfödd ensidig öronmissbildning och ledningsfel inkluderades i studien. Hörtrösklar för rena toner samt talförmåga i tyst och i brus mättes. Vidare fick försökspersonerna skatta sin hörselförmåga genom att besvara ett frågeformulär. Resultat: Resultaten visade försämrad hörselfunktion i det missbildade örat utöver ledningsfelet vid övertröskelig stimulering tydande på nedsatt funktion i innerörat eller i mer centrala delar av hörselsystemet. Vidare rapporterade försökspersonerna en mycket hög frekvens av hörselproblem i olika situationer.

Sammanfattande slutsatser

Studien visar att BAHA har likvärdiga långtidsresultat samt funktionsmönster hos både vuxna och barn talande för att BAHA även kan användas framgångsrikt på barn. Hos personer, både vuxna och barn med dubbelsidig hörselnedsättning visar resultaten bättre effekt av dubbelsidig BAHA jämfört med ensidig. Vid ensidig öronmissbildning och hörselnedsättning ses en klart försämrad hörselfunktion. I dessa fall kan ensidigt hörhjälpmedel vara av värde vilket testats hos barn med ensidig hörselnedsättning. Kompletterande större studier med utprovning av ensidig BAHA vid ensidigt ledningsfel samt dubbelsidig BAHA vid dubbelsidigt ledningsfel hos barn behöver göras innan generella behandlingslinjer kan utformas för dessa patientgrupper.