Does the Evidence Support Use of the Baha Implant System (Baha) in Patients with Congenital Unilateral Aural Atresia?

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Carole E. Johnson†
Melissa Mixon*

Abstract

Purpose: To determine if the evidence supports the recommendation of Baha implant systems (Bahas) over unaided conditions in persons with conductive hearing loss due to congenital unilateral aural atresia (CUAA), and if laboratory measures predict patient benefit and satisfaction.

Research Design: A systematic review.

Methods: The authors constructed and submitted search strings to PubMed and other electronic databases to identify studies in peer-reviewed journals that were at an appropriate level of evidence (systematic reviews, randomized controlled trials, or nonrandomized intervention studies); used outcome measures assessing audibility, localization, or speech-recognition in noise; included patients with CUAA using Bahas; and had interpretable data. References of all retrieved articles were also hand searched for relevant studies. Evaluation forms were completed by the authors for each of the included studies at all phases of the review including quality assessment and data extraction.

Results: The authors reviewed 88 retrieved titles and excluded four that had no relevance to the topic and 67 that were duplicates. Abstracts were reviewed for the remaining 17, and six nonrelevant studies were excluded. The remaining 11 articles were retrieved for full-text review; only three studies met inclusion criteria and were analyzed further. The three studies were not appropriate for a meta-analysis due to limited data, too few participants, and insufficient presentations of results. Qualitative analysis revealed inconsistent findings across audiometric measures, and few significant differences were noted with and without Bahas, yet most participants believed that Bahas improved their quality of life. Laboratory measures did not always predict patient benefit and satisfaction with Bahas.

Conclusions: Results were limited for this narrow population having CUAA and the specific criteria used for this review. Audiologic measures generally failed to predict patients’ success and/or satisfaction with their Bahas, but most of the included studies showed that patients perceived some benefits. Ideally, clinical decision making should include the highest levels of scientific evidence. However, when evidence is unavailable or does not support a clear-cut recommendation for a particular treatment across patients, as seems to be the case for the use of Bahas with CUAA, then clinicians must rely more heavily on clinical expertise and individual patient preferences in guiding clinical decision making.

Key Words: Baha implant system, congenital unilateral aural atresia, systematic review

Abbreviations: Baha = Baha implant system; CAA = congenital aural atresia; CONSORT = Consolidated Standards of Reporting Trials; CUAA = congenital unilateral aural atresia; EBP = evidence-based practice; HINT = Hearing in Noise Test; MAE = minimal average error; QoL = quality of life; SNR = signal-to-noise ratio; SRT = sentence recognition threshold

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Although most audiologists do not frequently encounter patients having congenital unilateral aural atresia (CUAA), it is still important for them to be prepared to be involved in the nonmedical diagnosis and treatment of these patients. Treatment options may include the Baha implant system (Baha). (Note that “Baha implant system” is used here rather than “bone-anchored hearing aid,” which was commonly used in the past.) Audiologists should be able to counsel those with CUAA and their families about current research findings, expert opinions, and previous patients’ experiences with various treatment options in order to help these patients make informed evidence-based practice (EBP) decisions.

Severity of congenital aural atresia (CAA) can vary from relatively minor cosmetic problems having little or no hearing loss to total absence of the ear canal, producing maximum conductive loss. CAA occurs in about 1/10,000–20,000 births. CUAA is about three times more common than bilateral atresia and is seen more often in males, frequently on the right side, and is usually sporadic. CUAA is sometimes due to genetic inheritance and can occur with craniofacial anomalies or syndromes (Kesser et al, 2008). CAA often presents reconstructive challenges resulting in many of these patients being poor candidates for surgical correction. Although surgery is still frequently used in CAA patients, increasingly, other options are being elected by those with CUAA (Niparko et al, 2003). Treatment options for CUAA and accompanying conductive hearing loss involve surgery, amplification including Bahas, or no intervention at all. Individuals with CUAA have difficulty localizing sounds, problems with the head shadow effect, and reduced speech recognition, especially in noise.

Intervention options, and their success, depend heavily on whether the atresia is unilateral or bilateral. Immediate amplification is essential for children with bilateral atresia because of the need for hearing to help develop speech and language. Also, surgery is not usually an option until children reach approximately five years. Although the need to intervene surgically is reduced significantly in children with CUAA when hearing is near normal in the nonaffected ear, achieving useable hearing from both ears as soon as possible is a major goal for children with atresia in order to prevent learning and communication delays.

Treatment options for CUAA with normal hearing in the unaffected ear have traditionally been left to the parents’ informed discretion. Some parents elect to do nothing on the atretic ear until their children are old enough to make their own decisions, while others initiate surgical reconstruction that often involves multiple procedures on the pinna with canal and middle ear reconstruction occurring much later. Recent data, however, have shown that there may be variable benefit relating to outcomes for speech perception in noise and binaural processing depending on the age at which surgery is initiated in patients with CUAA (Gray et al, 2009). Gray et al (2009) studied subjects with CUAA and tested their ability to understand speech in noise before and at 1 mo after having surgery to repair their atresia. Subjects were tested in a sound-treated booth with speech at 0 o azimuth and noise at 90 o to the side of the normal ear and again at 90 o to the atretic ear. All subjects were able to take advantage of a favorable signal-to-noise ratio (SNR) on the side of the newly opened ear (i.e., with noise toward the normal ear). Binaural squelch for understanding speech in noise directed to the atretic ear was exhibited in all but the youngest subjects under 8 yr of age. The authors stated that approximately 2 dB of binaural gain was lost for each decade that surgery was delayed, and they predicted that almost no binaural benefit would be noted after 38 yr of age, possibly due to longer periods of sensory deprivation.

Some families choose to use a traditional monaural air-conduction device on the better ear if it also has a hearing loss, or contralateral routing of the signal (CROS) or bone-conduction hearing aids, and/or Bahas on the atretic ear, sometimes with FM devices or amplified sound field in classrooms. A recent systematic review and meta-analysis showed that many children with unilateral hearing impairment have educational, behavioral, and/or speech and language delays (Cho Lieu, 2004), making the option of doing nothing less appropriate for many families and their hearing health-care providers. Further, as persons with CUAA mature, overall quality of life (QoL), levels of frustration, and issues related to self-efficacy for functioning in social situations, among other personal life experience factors, become increasingly important. These individual variables contribute to determine which type of treatment patients seek, and when, as well as their potential success.

The Baha is a surgically implantable osseointegrated system introduced in 1977 and approved for general use by the U.S. Food and Drug Administration in 1996 (Wazen et al, 1998). The Baha implant system consists of a titanium screw and external abutment, and sound processor that bypasses the outer and middle ears sending sound vibrations via the skull to the inner ear to promote hearing (Spitzer et al, 2002). The Baha might be a viable option for improving hearing in those with CUAA who may be inoperable or unsure about reconstructive surgery but still desire benefits of bilateral hearing (Niparko et al, 2003; Snik et al, 2004; Snik et al, 2005). The Baha has been shown to be safe and provide improvements in hearing and QoL for patients with unilateral sensorineural and bilateral conductive losses (Wazen et al, 1998; Newman et al, 2008). Successes, especially for improving audibility, localization, and
speech recognition in noise, have made the Baha an attractive option for potential use in adults with unilateral acquired conductive losses (Hol et al, 2005) and children (Spitzer et al, 2002). A soft headband can be used to permit children to derive benefits of the Baha in the interim before surgery (Spitzer et al, 2002). Further, Johnson et al’s (2006) systematic review found that patients with a wide variety of etiologies had reported improvements in QoL from Baha use.

Thus, audiolists, surgeons, and parents should be aware of the most current evidence available when deciding on treatment options for CUAA. Systematic reviews enhance EBP by formulating specific questions pertaining to certain diagnostic methods, medical treatments, and/or rehabilitative interventions; creating a priori criteria for study inclusion; and then amassing, evaluating, synthesizing, and interpreting relevant research evidence to answer the questions (Cochrane Collaboration, 2009). The purpose of this paper was to conduct a systematic review of the literature to address the following question: is there sufficient evidence (i.e., positive outcomes for sound localization, audibility, speech recognition in noise, and QoL) to support a recommendation for Baha use over no treatment (i.e., unaided) in patients with CUAA? The results of this review should provide information about whether there are consistent characteristics that lead patients to pursue the Baha as a treatment, audiological measures and self-assessment scales used to verify and/or validate Baha fittings, and benchmarks for success. The University of California Santa Barbara Institutional Review Board confirmed that human subjects were not involved in this systematic review.

METHODS

Search and Retrieval Strategy for Identification of Studies

The following summarizes the steps completed by the authors to conduct the systematic review. First, the authors framed the experimental question using the PICO rubric (Cox, 2005): population (CUAA), intervention (Baha), comparison (Bahas vs. unaided), and outcome (speech-in-noise, localization, audibility, or QoL).

The authors established a priori criteria to decide which studies to include/exclude in this systematic review. Included studies had to appear in an English-language, peer-reviewed journal (as determined by title and abstract reviews) and be at Evidence Level 3 or higher (as summarized in Table 1), involve patients with CUAA, and have interpretable data.

A full search strategy was used to identify studies that included data for patients with CUAA who were treated with Bahas. The search words and search string combinations shown in Table 2 were entered in the following databases: PubMed (including resources from 1950 to the present), ComDisDOME (2002 to 2008), CINAHL (Cumulative Index to Nursing and Allied-Health Literature, 1982 to the present), and Cochrane Database of Systematic Reviews (1952 to the present). The authors also hand searched all reference lists of retrieved articles for additional relevant studies.

After submitting the search words and search strings to the databases, the authors (1) inspected the lists of “hits” (i.e., titles of retrieved articles) and evaluated the titles, excluding those that clearly appeared to have no relevance to the research question for this study; (2) reviewed the abstracts of the included articles and assessed whether the criteria were completely satisfied (include), partially satisfied (unsure), or not at all satisfied (exclude); and (3) carefully analyzed the full text for the included and unsure abstracts, resulting in being classified as “include,” “exclude, but hold for potentially valuable information,” or “definitely exclude.” The authors independently evaluated each article and assessed its quality and were in 100% agreement regarding the articles to be included or excluded at each step.

Criteria for Included Studies

Study inclusion was assessed and recorded on a critical review sheet according to the following questions: (1) Was the Level of Evidence 3 or higher?; (2) Were outcome measures used including assessment of audibility, localization, speech recognition in noise, or QoL?; (3) Were participant groups clearly described as individuals with CUAA?; and (4) Were the data presented in numeric form that were clearly defined and relatable to patients with CUAA? The authors conducted a quality assessment of the included studies according to the criteria used by Chisolm and colleagues (2007). Data were extracted and qualitative information relevant to the posed question was summarized according to:

- Without Baha = With Baha (no evidence of statistically significant differences between outcome measures with or without Baha use)
- Without Baha > With Baha (evidence of statistically significant better performance on outcome measures unaided compared to with Baha use)

Table 1. Levels of Evidence Used for Rating Studies in This Systematic Review as Adapted from Cox (2005)

<table>
<thead>
<tr>
<th>Level Description of Evidence</th>
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<td>Systematic reviews and meta-analyses of randomized controlled trials (RCTs) or other high-quality studies</td>
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<td>3</td>
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<tr>
<td>Nonintervention studies: cohort studies, case-control studies, cross-sectional surveys</td>
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RESULTS AND DISCUSSION

The flow chart in Figure 1 summarizes the search and retrieval process that revealed that only three articles (i.e., Snik et al, 2002; Priwin et al, 2007a; Kunst et al, 2008) met criteria for inclusion. Eight studies (i.e., Wazen et al, 1998; Lustig et al, 2001; Wazen et al, 2001; Spitzer et al, 2002; Priwin et al, 2004; Snik et al, 2004; Wazen et al, 2007; Yellon, 2007) were excluded after full text review because they did not involve patients having CUAA. Additionally, some of these studies were not at an appropriate level of evidence and did not provide outcome measures meeting the criteria for this study or interpretable data. Quality assessment and summaries of the three included studies are provided in Tables 3 and 4, respectively.

Study Quality

The quality of the three studies was evaluated using the criteria of Chisolm and colleagues (2007), and the results are summarized in Table 3. All three studies were prospective audiometric evaluations at Evidence Level 3. Only Priwin et al (2007a) and Snik et al (2002) used control groups, but the data for the comparison conditions were questionable due to small sample size. Snik et al (2002) compared the performance of their patients to that of a control group with normal hearing, but few details were provided about the latter’s demographics. Priwin et al (2007a) was the only study to conduct a power analysis to determine values needed for an 80% chance of detecting a significant difference at the p < 0.05 level between and within groups for localization and speech recognition tasks. The other two studies used test-retest data from Snik et al (1998) to determine 95% confidence intervals for the magnitude of change in individual patient’s localization and speech recognition scores that would be necessary for significance at the p < 0.05 level.

Extensive details of patient inclusion and exclusion criteria were lacking in all three studies. However, all three studies adequately described their participant population regarding characteristics such as gender, age, time elapsed since implantation, pure-tone air/ bone-conduction thresholds, etiology of hearing loss, type of congenital anomaly, and hours per day and days per week of Baha use. Snik et al (2002) and Kunst et al (2008) reported more homogeneous grouping of patients than Priwin et al (2007a), whose sample included
several small subgroups having different types and degrees of hearing loss and treatment options. The procedures used for the surgery and fitting of the Baha were not reported in any of the studies, but general outcomes of surgery, existence of postoperative complications, and the health-care facility where the research was conducted were provided along with references. Only one study (Snik et al, 2002) provided information about the specific device that was fit, and none of the studies indicated whether omnidirectional or directional microphones were used. Finally, only Kunst et al (2008) discussed dropouts, but this was not critical given the pretest versus posttest group designs employed; omitting that information did not bias their results. Moreover, although the results for patients having CUAA were not well separated from those having other etiologies in most of the studies, the authors did report data from all patients regardless of amount of benefit achieved in audibility, localization, speech recognition, or QoL. The studies were not suitable for a meta-analysis due to small sample size.

### Participant Characteristics

All of the included studies were conducted in the Netherlands or Sweden, and the characteristics of the participants varied considerably. Only one study (Kunst et al, 2008) focused exclusively on the CUAA population. Snik et al (2002) used eight adults, but only two had CUAA. Kunst et al (2008) had the most homogeneous subject pool consisting of 20 CUAA participants (10 adults, 18–61 yr of age, and 10 children, 6–14 yr of age) with an average air-bone gap of 50 dB HL. Two of their subjects had congenital ossicular chain anomalies, which left only 18 with aural atresia. Priwin et al (2007a) had the most heterogeneous subjects consisting of 15 children with normal hearing (control group) and 22 having various forms of congenital unilateral or bilateral pinna, ear canal, and/or middle ear malformations. Unfortunately, the study included only four children with CUAA who used Baha, and two subjects had syndromes that confounded interpretation of the results. Thus, the three studies had very small numbers of subjects with CUAA and a wide range in ages (6–61 yr) and apparent individual differences.

### Outcome Measures

Generally, audiologic test paradigms for audibility, localization, and speech perception were consistent across the three studies. However, information relating to QoL was limited.

#### Audibility

Audibility using Baha versus unaided was generally measured by assessing subjects' aided and unaided hearing sensitivity for warble tones in sound field with (i.e., Snik et al, 2002; Kunst et al, 2008) or without (i.e., Priwin et al, 2007a) the better ear plugged. It is important to note that aided thresholds should come close to the bone-conduction thresholds of the better ear, but prefitting measures are via a transcutaneous bone-conduction vibrator while those for Baha are percutaneous through the implant, and one may not be an appropriate target for the other. Thus, there could be differences between the two, a variable that might affect target sound-field thresholds. All subjects having CUAA in these three studies demonstrated aided thresholds in the normal range regardless of whether the better ear was plugged or unplugged. These results were measured in laboratory conditions and may not accurately reflect benefits of the Baha when worn in “real-life” situations where the better ear would not be plugged, but one would assume that performance would be comparable.

#### Localization

Localization was generally assessed using five (Priwin et al, 2007a), seven (Snik et al, 2002), or nine (Kunst et al, 2008) loudspeakers situated in the horizontal plane in a half circle surrounding the subject. Subjects pointed to the loudspeaker delivering narrow-band noise-burst stimuli centered at either 500 and 2000 Hz (Snik et al, 2002) or 500 and 3000 Hz (Priwin et al, 2007a; Kunst et al, 2008) as shown in Table 4. All of the studies determined if each patient’s minimal average error (MAE in degrees) was significant. MAE was defined as the difference in azimuth (in degrees) between the origin of the stimuli and the subject’s perception of the signal, with smaller MAEs indicating better localization. The tasks were modified appropriately for children and adults.

All the studies showed wide variability in performance across listeners with CUAA. Some subjects’ performance improved with their Baha, others’ deteriorated, and a few reported no change. Snik et al (2002) explained that
Table 4. Summary of Studies Included

<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>Population</th>
<th>Outcome Measures and Results</th>
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</table>
| Kunst et al (2008)     | Prospective pre-post Baha audiometric evaluation | N = 20 consecutive patients with congenital unilateral conductive hearing loss and mean air-bone gap of 50 dB HL; 11, aged 18–61 yr, 9, aged 6–14 yr | Audibility: (unaided vs. aided) (Warble tones with and without Baha and the unaffected or normal ear plugged) 
Without Baha < With Baha for both adults and children |
|                        |                               | 18 had CUAA, 2 had congenital ossicular chain anomalies, All had Bahas (the particular models were not provided) | Sound Localization: (9 loudspeaker array in horizontal plane in 240° circle bow between −120° and 120° at 30° intervals; 1/3-octave noise centered at 500 or 3000 Hz at 65 dB SPL) 
Without Baha < With Baha |
|                        |                               |                                                                          | 3 of 10 patients for 500 Hz noise burst, 2 of 10 patients for 3000 Hz noise burst | 
Without Baha = With Baha | 5 of 10 patients for 500 Hz noise burst, 8 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | 2 of 10 patients for 500 Hz noise burst | 
Without Baha > With Baha | 2 of 10 patients for 500 Hz noise burst |
|                        |                               |                                                                          | Speech Recognition: (same as for adults except only five loudspeaker array at 60° intervals) | 
Without Baha < With Baha | 3 of 10 patients for 500 Hz noise burst, 2 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha = With Baha | 5 of 10 patients for 500 Hz noise burst, 8 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha > With Baha | 2 of 10 patients for 500 Hz noise burst |
|                        |                               |                                                                          | Children: | 
Without Baha < With Baha | 3 of 10 patients for 500 Hz noise burst, 2 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha = With Baha | 5 of 10 patients for 500 Hz noise burst, 8 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha > With Baha | 2 of 10 patients for 500 Hz noise burst |
|                        |                               |                                                                          | Speech Recognition: | 
Without Baha < With Baha | 3 of 10 patients for 500 Hz noise burst, 2 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha = With Baha | 5 of 10 patients for 500 Hz noise burst, 8 of 10 patients for 3000 Hz noise burst |
|                        |                               |                                                                          | Without Baha > With Baha | 2 of 10 patients for 500 Hz noise burst |
|                        |                               |                                                                          | In quiet (adaptive tracking procedure used to obtain SRTs to everyday Dutch sentences [Plomp and Mimpen, 1979]) | 
Without Baha < With Baha in | 4 of 10 patients | 
Without Baha = With Baha in | 6 of 10 patients | 
In noise and on side of the normal ear (adaptive tracking procedure used to obtain SRTs to everyday Dutch sentences with speech in front via a loudspeaker and steady-state noise with same spectrum as the sentences was fixed at 65 dB SPL via loudspeaker on left or right of subject) | 
Without Baha < With Baha in | 4 of 10 patients | 
Without Baha = With Baha in | 6 of 10 patients | 
In noise and on side with Baha |
<table>
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<th>Outcome Measures and Results</th>
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<tr>
<td>Priwin et al (2007a)</td>
<td>Prospective pre-post Baha audiometric evaluation</td>
<td>N = 22 children with either single-sided or bilateral conductive hearing loss&lt;br&gt;15 control subjects&lt;br&gt;9 girls, 13 boys&lt;br&gt;Age = 6–17 yr&lt;br&gt;21 had congenital malformations&lt;br&gt;13 had isolated malformations&lt;br&gt;7 had syndromes&lt;br&gt;7 had unilateral conductive losses and no amplification</td>
<td>Audibility: (unaided vs. aided)&lt;br&gt;Measured in sound field using warble tones through a loudspeaker at 0° azimuth with and without Baha and no plug in unaffected or normal ear&lt;br&gt;Without Baha = With Baha&lt;br&gt;Sound Localization: (Five loudspeaker array in horizontal plane in a frontal semicircle; narrow-band 1/3-octave noise centered at 500 or 3000 Hz at 50 and 60 dB SPL)&lt;br&gt;Without Baha = With Baha and in some cases:&lt;br&gt;Without Baha &gt; With Baha for 500 and 3000 Hz noise bursts&lt;br&gt;Sound localization very poor and close to chance for single-sided Baha use&lt;br&gt;Speech Recognition:&lt;br&gt;Speech (10 lists of phonemically balanced Swedish three-word sentences [extracted from Hagerman, 1982]) in noise (speech was fixed at 60 dB SPL and noise was at 0, 4, and 6 dB SNRs)&lt;br&gt;Without Baha = With Baha</td>
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Table 4. Continued

<table>
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</tr>
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<tbody>
<tr>
<td>Snik et al (2002)</td>
<td>Prospective pre-post Baha audiometric evaluation</td>
<td>N = 8 adults with inoperable unilateral air-bone gaps</td>
<td>Audibility: (unaided vs. aided) &lt;br&gt;Measured in sound field using warble tones with and without Baha and the “subnormal” ear plugged</td>
</tr>
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<td></td>
<td></td>
<td>2 had CUAA</td>
<td>Sound Localization: (Seven loudspeaker array in horizontal plane in a half circle at 30° intervals; narrow-band noise centered at 500 or 2000Hz at 60 dB SPL)</td>
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<tr>
<td></td>
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<td>Age = 19–51 yr</td>
<td>Without Baha = With Baha for both 500 and 2000Hz noise burst; unaided localization abilities in patients with CUAA well developed</td>
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<tr>
<td></td>
<td></td>
<td>3 had high-frequency sensorineural hearing loss and unilateral air-bone gap</td>
<td>Speech Recognition: &lt;br&gt;In quiet (details were not provided) &lt;br&gt;Without Baha = With Baha &lt;br&gt;One patient with CUAA had significant improvement; the other one did not</td>
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<td>Subjects were fitted with the Baha Classic HC300 (Entific, Gothenburg, Sweden)</td>
<td>In noise and on side of normal ear &lt;br&gt;(adaptive tracking procedure used to obtain SRTs to everyday Dutch sentences [Plomp and Mimpen, 1979] with speech in front via a loudspeaker and noise fixed at 65 dB SPL via loudspeaker on left or right of subject)</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Without Baha = With Baha &lt;br&gt;One patient with CUAA had improvement in SRT with noise on side of normal ear; the other one did not</td>
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</table>

QoL: <br>Without Baha < With Baha based on two patients’ daily use and being happy with their devices

Note: CUAA = congenital unilateral aural atresia; IOI-HA = International Outcome Inventory for Hearing Aids; QoL = quality of life; SNR = signal-to-noise ratio; SRT = sentence recognition threshold.
some subjects with CUAA had remarkably good unaided localization abilities and had learned coping strategies, which may have obscured benefits provided by the Baha in these laboratory conditions.

Speech Recognition

Speech recognition performance in noise can be problematic for patients with unilateral hearing loss, particularly when the noise source is positioned near the better ear and the speech stimulus is presented at or near the poorer ear. Ideally, wearing a Baha on the affected side would improve that situation. It could be hypothesized, however, that some patients with unilateral air-bone gaps may experience deterioration in speech recognition performance due to possible interference caused by amplification of the noise when presented on the same side as the Baha. The testing paradigms used by Snik et al (2002) and Kunst et al (2008) attempted to create these situations in the laboratory environment to assess the benefits of Bahas for improving speech recognition (as well as degradation).

Speech recognition was assessed differently for adults and children across the studies. Snik et al (2002) determined the SNRs needed for adults to reach 50% everyday Dutch sentence recognition thresholds (SRTs) using an adaptive procedure both in quiet and using speech-shaped noise via two loudspeakers. One loudspeaker presented the sentences at a 0° azimuth at varied sound pressure levels (SPLs). The other presented the noise at 65 dB SPL to either the right or left of the subject. Resulting SRTs were the average of two trials in each condition. Kunst et al (2008) used a similar procedure but measured phoneme recognition for the children using common Dutch words presented at 60 dB SPL at 0° azimuth via a loudspeaker with noise delivered to the side of the child’s best ear at 65 dB SPL through another loudspeaker creating a fixed –5 dB SNR. Similarly, Priwin et al (2007a) measured sentence recognition scores using phonemically balanced lists of three-word Swedish sentences presented at 60 dB SPL with background noise via the same loudspeaker at 0° azimuth adjusted to three different levels of 60, 56, and 54 dB SPL creating SNRs of 0, +4, and +6 dB, respectively.

Speech recognition in noise was variable for subjects with CUAA as reflected by the results for the two adults in the Snik et al (2002) study. The authors reported that one of the subject’s scores were significantly improved while the other had no change in the SNR score. Kunst et al (2008) found significant improvement in speech recognition thresholds from unaided to aided conditions for five of eight children for whom complete data sets were available. It is not known, however, whether the results were significant for their entire group because unaided speech recognition scores were not available for two of the 10 children. The authors indicated that subjects’ consistent use of Bahas was highly suggestive of benefit and that trial periods with soft headbands might help to evaluate whether patients are suitable candidates for Bahas. Additionally, Priwin et al (2007a) found mean improvements in children’s Baha over unaided performance at the most difficult of the three SNRs (i.e., 0 dB), but not for the easier (i.e., +4 and +6dB) SNRs. Although individual subject results were not presented, the limited findings from these studies showed promise that use of the Baha, particularly for children, may improve speech recognition ability in noise.

In summary, as was seen for localization results, the amount of benefit provided by the Baha for speech recognition in quiet and in noise was inconsistent across the three studies. The benefits achieved by adults were not as consistent as those reported for the children. Even for children, however, the number of patients was too small to determine any definitive trends in performance, and differences between age groups may have been due to the sensitivity of the speech recognition tasks used. Interestingly, as noted earlier, Gray and colleagues (2009) found similar inconsistencies for speech recognition and binaural performance in patients with CUAA who underwent surgery at different ages. The authors suggested that little benefit might be gained from surgery after approximately 38 yr of age due to possible effects of sensory deprivation, and that performance in children younger than 8 yr was poorer but might improve over time. Thus, these are important issues for families deciding whether to pursue surgery or Bahas as treatment options.

A positive and consistent finding across the studies was that when noise was presented on the same side as the Baha, the performance by most subjects was not negatively affected. This may help alleviate the concern that amplification of noise from the Baha would cause interference for listeners.

Quality of Life

Many types of standardized and informal instruments have been used by audiologists to assess patients’ QoL (Chisolm et al, 2007), but the measures used in the three studies were minimal, and the authors alluded that the Baha provided improvement in the subjects’ QoL. Priwin et al (2007a) used a validated Swedish version of the seven-item International Outcome Inventory for Hearing Aids (IOI-HA; Cox et al, 2002) for their children’s (or parents’ when necessary) self-assessments of their QoL with the Baha. Even so, only item seven directly asked about QoL and suggested an improvement with amplification, but unfortunately, the authors did not segregate the data for children using hearing aids from those having Bahas, which confounded...
interpretation of the results. The other two studies only inferred improvements in QoL by reporting on subjects’ compliance with and daily use of their Baha. Kunst et al (2008) did not measure QoL but indicated that their subjects’ consistent use and high compliance with their Baha was suggestive of benefit. Likewise, although Snik et al (2002) did not assess QoL, these researchers indicated that no adverse effects were reported for their two subjects with CUAA and that even though their audiologic improvements with their Baha was limited, the subjects used their Baha daily and were pleased with performance, which was interpreted as implying improvement in QoL.

Kunst et al (2008) was the only study that included a reasonable sample of 18 adults and children with CUAA fit with Baha. Clearly, additional studies are needed to support the recommendation that use of Baha increases audibility and QoL in patients with CUAA.

Clinical Implications

Ultimately, the amount of evidence retrieved in this systematic review was not adequate for making conclusive recommendations for patients and their families considering Bahas as treatment options for CUAA. One certainty was that the contribution from the Baha for localization and speech recognition ability was highly variable between subjects; some improved, others stayed the same, and a few deteriorated. There may be several reasons for this, and further research is needed to determine which subject factors best predict improvement in auditory skills through Baha with CUAA.

One possible prognostic patient characteristic factor may be the magnitude of pretreatment air-bone gap. Hol et al (2005) and Priwin et al (2007b) stated that subjects with unilateral conductive hearing loss who benefited the most from Baha were those with air-bone gaps of 50 dB HL or greater. Critical periods for the development of localization skills and auditory deprivation are other factors that may help predict the benefit provided by Baha fitted to older children and adults with CUAA. For example, Gray et al (2009) suggested that the critical period for these skills might be by the end of the third decade of life based on subjects who received surgery to correct their CUAA. Some patients who are well beyond the critical period for developing those skills may simply not be able to use cues for localization provided by a Baha, at least without additional training and/or exposure. Alternatively, the results from the included studies suggest that some subjects with CUAA may have already developed compensatory skills for localization in real-world listening environments that prevented the experimenters from documenting improvements with their Baha when compared to the unaided conditions. Another reason for variable performance noted both within and across the studies was that subjects differed in the amount of time that they used the Baha (Snik et al, 2002). Yet another reason was that in some subjects, the hearing in the better ear was near normal, and minimal benefit was obtained from the Baha. Clearly, more research is needed on the effects of the Baha on the auditory skills of subjects with CUAA. In the interim, as recommended by Priwin et al (2007a, 2007b), use of a Baha Softband during a trial period may provide some indication of expected benefit. In this way, patients and their families could weigh the potential benefits before undergoing surgery.

The results of this systematic review have implications for audiologists counseling patients with CUAA and their families about possible treatment options including Baha. No consistent commonalities were found from the three studies as to why patients with CUAA should pursue Baha, at least from the subjects’ performance in Baha and unaided conditions. The small number and heterogeneous nature of the patients with CUAA precluded establishing candidacy criteria that could be used to predict success with Baha.

All three studies were fairly consistent in their use of audiologic (laboratory) and self-assessment QoL measures. The audiologic measures attempted to assess benefits of binaural hearing usually thought to be unavailable to patients with unilateral hearing loss. Some patients with CUAA, however, apparently had already developed localizing coping strategies as evidenced by their normal unaided performance (Snik et al, 2002). For whatever reason, localization results from the controlled laboratory conditions in these studies using multiple loudspeaker arrays in an arc around the listener with and without the Baha provided little information for predicting patient outcome. Although many audiologists do not have the equipment or time to conduct localization measures in busy clinical practices, clinicians do have the skills and equipment to assess improvements in speech recognition in quiet and noise for patients with unilateral hearing loss. Audiologists can simulate the paradigms used in these studies with instruments such as the Hearing in Noise Test (HINT; Nilsson et al, 1994) or the Quick Speech-in-Noise (QuickSIN; Killion et al, 2004). Audiologists should be advised, however, that, as can be said for any amplification system, results gleaned from such measures may not necessarily predict real-world performance with Bahas in patients with CUAA. Similarly, audiologists should remember that although comparing unaided thresholds or those obtained under earphones to aided performance with the normal ear plugged may be used to verify functional gain of the Baha on the affected side, these measures do not necessarily infer similar real-world benefit because of normal hearing sensitivity in the unaffected ear. Audiologists should not really be interested in functional gain but, rather,
if the aided threshold agreed with the bone-conduction threshold of the better ear. Therefore, there is little evidence available regarding the utility of these measures in clinical situations, and audiologists should not use these test results alone for assessing patients’ candidacy for, or predicting their success with, Baha in the real world. It has been hypothesized that relatively good unaided localization behaviors in some patients with CUAA might suggest that they have developed adequate compensatory strategies. Neither adequate pretreatment performance nor negligible postfit gains, however, necessarily suggest that a given patient with CUAA will or has not benefited in this skill from a Baha. Indeed, it was reported in these studies that regardless of whether improvement with Baha was achieved on objective laboratory measures, nearly all of the subjects used their devices during waking hours most days of the week. Compliance has been suggested to be an indicator of patients’ satisfaction, and the limited evidence recovered here showed that use of and benefits derived from Baha may not be predicted from performance on audiological measures alone.

So, what clinical expectations can audiologists be sure of with little or no evidence about these patients and devices? Audiologists can be reasonably assured that the Baha is a low-risk treatment for patients with CUAA that has potential real-world benefits, particularly for those having 50 dB air/bone gaps on the affected side. The lack of any serious complications from the Baha indicates it is unlikely that patients will suffer adverse affects from the Baha. Indeed, the most consistent finding was that for adults with CUAA, use of the Baha did not result in deterioration of performance when noise was on the side with the device and speech was delivered at the normal ear. Thus, audiologists can counsel patients that speech recognition performance in noise should not decrease using a Baha. For patients who may be anxious about the surgery to implant the device, a Baha Softband can provide a reasonable trial period before committing to implantation.

Some audiologists have become accustomed to using EBP in counseling patients and their families weighing the advantages and disadvantages of various treatment options. Jerger (2008) commented on Olson and Shinn’s (2008) systematic review of the effectiveness of using amplification with cochlear implants. He aptly noted that researchers’ desires to embrace EBP from studies involving large groups of subjects is not always possible and may not result in answers that satisfy clinicians who need information about specific patients. Indeed, individual differences do confound EBP, especially when questions arise about patients having peculiar pathologies like CUAA that may not lend themselves to investigations where large numbers of subjects are possible. Wisely, the investigators in two (Snik et al, 2002; Kunst et al, 2008) of the three studies determined confidence intervals for laboratory measures that would indicate a significant change in performance for subjects at the $p < 0.05$ level.

The overall dilemma resulting from the present study was similar to that of Olson and Shinn (2008), who found few quality articles at a high level of evidence relating to subjects’ bimodal use of hearing aids with cochlear implants. Similar to the individual differences Olson and Shinn (2008) found in subjects across studies, the authors of this systematic review observed that individual subjects in the included studies used and were happy with their Baha in spite of the fact that subjects did not always achieve improved test scores in laboratory conditions with use of the Baha. The authors agree with Jerger (2008) that research focusing on large-sample studies that measure success by whether subjects’ improvement with their devices meets statistical significance may not always be practical for answering clinical questions about who does or does not benefit from a specific technology, at least in small populations like CUAA. Rather, audiologists might emphasize the importance of outcome measures that are highly individualistic, such as the Client-Oriented Scale of Improvement (COSI; Dillon et al, 1997) that has patients generate and prioritize listening situations to target for improvement in auditory rehabilitation. For children, input from parents and teachers is particularly critical, and in such cases, the Client-Oriented Scale of Improvement for Children (COSI-C; National Acoustic Laboratories, 2009) may be useful. Individualizing treatment goals is necessary when little or no evidence is available to provide estimates of the likelihood of success with treatment for relatively rare conditions like CUAA that affect patients and their families in unique ways. Thus, clinicians may be forced to rely on patients’ impressions about the appropriateness and benefits of Bahas, particularly when little or no scientific evidence is available for clinical decision making.

**CONCLUSIONS**

Overall, this review provided limited evidence for a strong recommendation supporting the use of Baha in patients with CUAA. The small number of subjects having CUAA in each study was a major limitation (Snik et al, 2002), especially when results for those with different etiologies were combined into a single data set. Although the Baha has been reported to be quite beneficial for restoring binaural hearing in patients having other etiologies, those with CUAA must learn to cope with a variety of cosmetic and functional hearing ability obstacles early in life. Many patients with CUAA and near-normal or corrected hearing in the nonaffected ear seem to compensate quite well in the conditions evaluated in these studies, sometimes even without Baha, which made it difficult to assess the benefit of
the Baha in laboratory environments. Nevertheless, many Baha users reported satisfaction with the Baha and preferred to use the Baha regularly. This suggested that the outcome measures employed in the laboratory controlled situations of these studies may not be sensitive enough to detect improvements with the Baha that patients might realize in real-world use. Out of necessity, many subjects with CUAA appeared to have developed compensatory auditory skills and had excellent monaural unaided localization abilities.

Therefore, audiologists should consider a paradigm shift in reviewing treatment options for patients with CUAA. Instead of reviewing the success of the Baha with this population as a whole, a patient-centered approach focusing on personal history and communication needs might be more effective for determining the appropriateness of the Baha for individual patients. To their credit, the authors of the studies had the foresight to conduct power analyses (Priwin et al., 2007a) or use confidence intervals (Snik et al., 2002; Kunst et al., 2008) to calculate performance increments required to achieve statistically significant differences at the level of the patient. In other words, these authors recognized the high degree of intersubject variability that is common in this type of study and considered the significance of unaided versus aided performance for each subject rather than focusing on group data. Future research should collect individual subject data from various facilities throughout the world using the consolidated standards of reporting trials (CONSORT) format and aggregate the findings in national and/or global repositories to determine optimal patient selection criteria and expected outcomes for Baha with CUAA (Moher et al., 2001). The 22-item CONSORT checklist can assist researchers in providing the complete and accurate information necessary for publishing controlled clinical trials and should facilitate comparisons across research sites, subjects, and technologies. Further, the use outcome measures at the end of an experiment, so-called surrogate endpoints, do not guarantee the long-term benefit of Bahamas over the months and years ahead. In addition, the literature is replete with studies using objective laboratory and subjective questionnaire measures of QoL showing the benefits of Baha in subjects with sensorineural and bilateral conductive hearing loss. In the studies reviewed here, however, subjects with CUAA displaying minimal benefit on laboratory measures were satisfied with the Baha and reported a reduction in activity limitations when using the Baha. The lack of evidence supporting a strong recommendation for use of the Baha in patients with CUAA reported from this systematic review should not discourage researchers from continuing to assess improvement. The laboratory measures and data-reporting methods used may simply need to be refined. Likewise, prospective patients and their families should not be discouraged from considering the Baha as a treatment option, especially considering that noninvasive Softband trials can be conducted before deciding to pursue implantation.

Currently, audiologists must counsel their patients that there is little evidence at the most rigorous levels in peer-reviewed literature to support the use of Baha for CUAA. Audiologists should know that although this review searched databases from the 1950s to the present, realistically, the Baha has only been available for general use since 1996 (Wazen et al., 1998), and its consideration as a treatment for CUAA has only occurred in the past decade. This and the comparatively small number of patients with CUAA led to few studies being found in the literature that met the inclusion criteria. Until high-level EBP studies become available, audiologists, patients, and their families may rely on other forms of evidence including expert opinion, results from previous users, and patients’ testimonials in making the decision about whether to pursue Baha for CUAA.

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REFERENCES


