Клинические сведения

Атрезия/микротия и Baha®

Система Cochlear™ Baha® 3

Врожденная атрезия наружного слухового прохода – это отсутствие наружного слухового прохода при рождении. Этот порок часто сопровождается микротией – пороком развития наружного уха с деформацией ушной раковины. Тяжесть микротии варьирует от незначительной деформации ушной раковины до полного отсутствия наружного уха. В таких случаях очень важно раннее слухопротезирование, чтобы у ребенка не было задержки языкового и речевого развития. Опубликованы великолепные результаты применения системы Baha у детей с атрезией слухового прохода с подробными описаниями успешных случаев[13-16].

Атрезия/микротия возникают в результате нарушений развития жаберных дуг у эмбриона. Они также могут быть одним из проявлений более сложных генетических синдромов, в том числе синдрома Гольденхара и синдрома Тричера-Коллинза. При выборе наилучшего метода лечения при атрезии очень важно оценить степень тяжести данного порока развития. Для этой цели предложено несколько балльных шкал, из которых наиболее часто используется шкала оценки степени тяжести Ярсдерфера (Jahrsdoerfer)[1,2]. Эта шкала включает ряд параметров, каждому из которых присвоено определенное количество баллов, и общая сумма баллов помогает принять решение о наилучшем методе лечения для конкретного ребенка.

ЧАСТОТА

По разным источникам, атрезия слухового прохода встречается с частотой от 1/10000 до 1/20000 новорожденных[3,4]. В 85% случаях наблюдается односторонняя атрезия слухового прохода[4]. Атрезия слухового прохода более чем в 2 раза чаще встречается у мальчиков, чем у девочек[3]. Когда атрезия слухового прохода является одним из проявлений генетического синдрома, двустороннее поражение наблюдается намного чаще, в 50% случаев[4].

ЗАДЕРЖКА РАЗВИТИЯ

Двусторонняя атрезия слуховых проходов без слухопротезирования проводит к задержке речевого и когнитивного развития.
Детям с двусторонней атрезией рекомендуется двустороннее слухопротезирование, начиная с младенческого возраста. Результаты все большего числа исследований свидетельствуют о важности раннего вмешательства не только при двусторонней, но и при односторонней тугоухости[5–7]. По данным ряда исследований, у детей с односторонней атрезией слухового прохода отмечается задержка развития речи и часто возникают трудности при обучении в школе[6,8]. Кроме того, у таких детей чаще возникают социальные, поведенческие и психологические проблемы, чем у детей с нормальным слухом[9]. В дополнение к вышесказанному следует отметить, что у детей с врожденной односторонней тугоухостью позднее слухопротезирование не приносит ощутимой пользы в отношении восстановления бинаурального слуха, возможно, это объясняется тем, что слуховой аппарат человека не способен в полной мере использовать все возможности слухопротезирования, если в ранние годы развитие слуха было нарушено[10]. При таких заболеваниях, как односторонняя атрезия слухового прохода, необходимо предупреждать появление возможных проблем в будущем; для этого следует как можно раньше информировать родителей о риске задержки развития и возможных вариантах решения этой проблемы.

ВОССТАНОВЛЕНИЕ СЛУХА С ПОМОЩЬЮ BAHA

Восстановление слуха приоритетнее устранения косметических дефектов, поскольку сводит к минимуму риск задержки речевого и когнитивного развития. Согласно результатам нескольких исследований, ношение звукового процессора Baha на эластичной ленте с младенческого возраста способствует устранению задержки развития речи[11,12]. При двусторонней атрезии рекомендуется двустороннее слухопротезирование, чтобы ребенок мог пользоваться всеми преимуществами бинаурального слуха, такими как локализация источника звука, бинауральная суммация громкости и более четкое различение звуков в шумной обстановке[13]. Детям старшего возраста для успешного восстановления слуха может потребоваться установка имплантата Baha. Для определения оптимального времени имплантации полезно следить за темпом речевого развития ребенка. Речевое развитие может замедлиться, когда ребенок станет старше и его речь будет более развитой. Если у ребенка отмечается замедление речевого развития, рекомендуется регулярно его оценивать и в скором времени установить имплантат Baha; успешность этого подхода была доказана[12].

У детей, у которых в более старшем возрасте не исключается проведение хирургической реконструкции наружного уха, очень важно правильно выбрать место установки имплантата Baha. Возможно, потребуется расположить имплантат несколько кзади от стандартного места имплантации для сохранения возможности проведения успешной реконструктивной операции, однако не слишком далеко, чтобы не ухудшить качество слуха. Согласно текущим рекомендациям, расстояние от имплантата до наружного отверстия слухового прохода должно составлять не менее 60–65 мм. Если реконструкция будет успешной, находящуюся в толще кожи опору Baha можно будет легко удалить. Если же хирургическое вмешательство будет неудачным, опору можно вновь подсоединить к имплантату и вернуться к использованию звукового процессора для восстановления слуха.

ПРЕИМУЩЕСТВА BAHA

Основные преимущества применения системы Baha при атрезии слухового прохода:

- С помощью системы Baha можно безопасно и эффективно восстановить слух у детей с односторонней и двусторонней атрезией слухового прохода, начиная с младенческого возраста[12,13].
- Применение системы Baha дает великолепные аудиологические результаты у детей как при установке, так и впоследствии[14,15].
- У пациентов, пользующихся системой Baha, отмечаются наиболее высокие уровни удовлетворенности по шкале Glasgow Benefit по сравнению с другими подгруппами[15].
- В случаях односторонней атрезии слухового прохода установку системы Baha следует рассматривать как способ уменьшения риска задержки развития и нарушения формирования навыков общения[16].
- По сравнению с хирургической реконструкцией установка системы Baha представляет собой экономически оправданный, безопасный и эффективный способ восстановления слуха с превосходными аудиологическими результатами[17].

ЗАКЛЮЧЕНИЕ

Получено множество научных доказательств, свидетельствующих о великолепных результатах применения системы Baha при атрезии слухового прохода. Раннее слухопротезирование крайне важно как при двусторонней, так и при односторонней атрезии[4–6]. Опубликованы великолепные результаты применения системы Baha у детей с атрезией слухового прохода – отмечались не только превосходные аудиологические результаты, но и высокие уровни удовлетворенности пациентов, использующих систему Baha[13-16].

СПИСОК ЛИТЕРАТУРЫ

1. Jahrsdoerfer RA, Yeakley JW, Aguilar EA, Cole RR, Gray LC. Grading system for the selection of patients with congenital aural atresia. The American journal of otology. 1992 Jan;13(1):6-12.

ABSTRACT
It is generally recognized that surgery for congenital aural atresia is difficult. In an effort to select those patients who have the greatest chance of success, we have developed a grading scheme based on the preoperative temporal bone CT scan and the appearance of the external ear. Patients are graded on a possible best score of 10. The stapes is assigned the highest rating (2 points), while all other entrees on the scale are 1 point. The grade assigned preoperatively has been shown to correlate well with the patient's chance of success, herein defined as a postoperative speech reception threshold of 15 to 25 dB. A patient with a preoperative grade of 8/10 would, therefore, have a 80 percent chance of achieving this threshold. Patients with scores of 5/10, or less, are not considered surgical candidates, because the risk of the operation would outweigh the potential benefits. We have found that the grading system allows us to avoid impossible surgical cases while allowing for a reasonable prediction of the hearing outcome.

2. Shonka DC Jr, Livingston WJ 3rd, Kesser BW. The Jahrsdoerfer grading scale in surgery to repair congenital aural atresia. Archives of otolaryngol ogy head & neck surgery. 2008 Aug;134(8):873-7.

ABSTRACT
OBJECTIVE: To determine the predictive ability of the Jahrsdoerfer grading scale score in congenital aural atresia surgery.
DESIGN: Retrospective review of medical records.
SETTING: Tertiary referral center.
PATIENTS: One hundred eight patients with aural atresia.
MAIN OUTCOME MEASURES: Demographic data, preoperative Jahrsdoerfer score, and postoperative audiometric outcomes were reviewed. One month postoperative, 4-tone pure-tone averages and speech reception thresholds were compared between ears scoring 6 or lower, 7, and 8 or higher on the Jahrsdoerfer grading scale. The percentage of ears with a speech reception threshold of 30 dB hearing level or lower for each group was calculated and compared between groups. Individual anatomical structures on the Jahrsdoerfer grading scale were evaluated for their ability to predict postoperative audiometric success.
RESULTS: Of 116 ears evaluated, postoperative 4-tone pure-tone averages and speech reception thresholds were significantly poorer in ears scoring 6 or less on the Jahrsdoerfer grading scale compared with ears scoring 7 or higher ($P < .02$, t test). Ears scoring 6 or less had a 45% chance of achieving a postoperative speech reception threshold of 30 dB hearing level or lower, while ears scoring 7 or higher had an 89% chance ($P < .01$, chi(2) test). Lack of middle ear aeration was the only anatomical factor predictive of poor audiometric outcome.
CONCLUSIONS: Compared with patients with a Jahrsdoerfer score of 6 or lower, patients with a score of 7 or higher had significantly better hearing postoperatively. Middle ear aeration may be the most important predictor of postoperative hearing outcome. The Jahrsdoerfer grading scale is an invaluable tool in the preoperative evaluation of patients with congenital aural atresia.

3. Kelley PE, Scholes MA. Microtia and congenital aural atresia. Otolaryngologic clinics of North America. 2007;40:61-80.

ABSTRACT

Congenital aural atresia (CAA) and microtia are congenital anomalies that are so common that every otolaryngologist should be familiar with the initial evaluation and care of the patient. When one ear hears normally, speech and language development should be normal. The gross and fine motor development of the baby or child is not expected to be affected in isolated cases of microtia and CAA. Current technologies allow for reconstruction or habilitation of the microtic ear when the child is several years of age. The hope is that tissue engineering can eliminate donor site morbidity. Temporary prosthetic ears will remain an option. Aural atresia work continues to be very dependent on the patient anatomy and the need or desire for better hearing in the affected ear.

4. Mastroiacovo P, Corchia C, Botto LD, Lanni R, Zampino G, Fusco D. Epidemiology and genetics of microtia-anotia: a registry based study on over one million births. Journal of medical genetics. 1995;32(6):453-7.

ABSTRACT

The epidemiology and genetics of microtia-anotia (M-A) were studied using data collected from the Italian Multicentre Birth Defects Registry (IPIMC) from 1983 to 1992. Among 1,173, 794 births, we identified 172 with M-A, a rate of 1.46/10,000; 38 infants (22.1%) had anotia. Of the 172 infants, 114 (66.2%) had an isolated defect, 48 (27.9%) were multiformed infants (MMI) with M-A, and 10 (5.8%) had a well defined syndrome. The frequency of bilateral defects among non-syndromic cases was 12% compared to 50% of syndromic cases ($p = 0.007$). Among the MMI only holoprosencephaly was preferentially associated with M-A (four cases observed upsilon 0.7 expected, $p = 0.005$). No significant variations were identified in the prevalence of non-syndromic cases by geographical area (range 0.62-2.37/10,000 births) or by five month time periods (range 0.21-2.58/10,000 births), nor was there evidence of time trends. When M-A cases were compared to controls, we found that mothers with parity 1 had a higher risk of giving birth to an MMI with M-A, and that mothers with chronic maternal insulin dependent diabetes were at significantly higher risk for having a child with M-A. MMI with M-A had higher rates of prematurity, low birth weight, reduced intrauterine growth, and neonatal mortality than infants with isolated M-A and controls. Babies with isolated M-A had, on average, a lower birth weight than controls; the difference was higher for females. The analysis of pedigrees and familial cases suggests an autosomal dominant trait with variable expression and incomplete penetrance in a proportion of cases, or a multifactorial aetiology. Three cases had consanguineous parents, but the absence of M-A among previous sibs does not support autosomal recessive inheritance.

5.Bess F. H., Tharpe A. M., and Gibler A. M.: Case history data on unilaterally impaired children. Ear and hearing. 1986, 7, 1; 14-19.

ABSTRACT

This paper presents the data from medical and educational case histories on a group of 60 unilaterally hearing-impaired children. The case history data revealed that approximately one-half of the 60 children with unilateral sensorineural hearing loss exhibited some difficulty in educational progress. More specifically, 35% had failed at least one grade and an additional 13% were in need of some special resource assistance. Similar findings were obtained on a subset of 25 unilaterally hearing-impaired children who satisfied rather stringent criteria for age, hearing level, intelligence, length of time the impairment was present, history of middle ear disease, and general growth and development.

6. Lieu JEC, Tye-Murray N, Karzon RK, Piccirillo JF. Unilateral hearing loss is associated with worse speech language scores in children. Pediatrics. 2010 Jun;125(6):e1348-e1355.

ABSTRACT

OBJECTIVE: To determine whether children with unilateral hearing loss (UHL) demonstrate worse language skills than their siblings with normal hearing, and whether children with UHL are more likely to receive extra assistance at school.

PATIENTS AND METHODS: We conducted a case-control study of 6- to 12-year-old children with UHL compared with sibling controls (74 pairs, n = 148). Scores on the oral portion of the Oral and Written Language Scales (OWLS) were the primary outcome measure. Multivariable analysis was used to determine whether UHL independently predicted OWLS scores after we controlled for potential confounding variables.

RESULTS: Children with UHL had worse scores than their siblings on language comprehension (91 vs 98; P = .003), oral expression (94 vs 101; P = .007), and oral composite (90 vs 99; P < .001). UHL independently predicted these OWLS scores when multivariable regression was used with moderate effect sizes of 0.3 to 0.7. Family income and maternal education were also independent predictors of oral expression and oral composite scores. No differences were found between children with right- or left-ear UHL or with varying severity of hearing loss. Children with UHL were more likely to have an individualized education plan (odds ratio: 4.4 [95% confidence interval: 2.0-9.5]) and to have received speech-language therapy (odds ratio: 2.6 [95% confidence interval: 1.3-5.4]).

CONCLUSIONS: School-aged children with UHL demonstrated worse oral language scores than did their siblings with normal hearing. These findings suggest that the common practice of withholding hearing-related accommodations from children with UHL should be reconsidered and studied, and that parents and educators should be informed about the deleterious effects of UHL on oral language skills.

7. Keogh T, Kei J, Driscoll C, Khan A. Children with Minimal Conductive Hearing Impairment: Speech Comprehension in Noise. Audiology & neuro-otology. 2010;15:27-35.

ABSTRACT

Based on a study sample of 1071 primary school children (5.3-11.7 years), 10.2% of the children were found to have conductive hearing loss in 1 or both ears. Binaural speech comprehension scores of a subset of 540 children were analyzed. The results showed that children with bilateral conductive hearing loss had the lowest mean scores of 60.8-69.3% obtained under noise conditions. These scores were significantly lower than the corresponding scores of 69.3-75.3% obtained by children with possible middle ear disorders but no apparent hearing loss, 70.5-76.5% obtained by children with a unilateral conductive hearing loss and 72.0-80.3% obtained by their normally hearing peers. This study confirms that young children, who are known to have poorer speech understanding in noise, show further disadvantage when a bilateral conductive hearing loss is present.

8. Lieu JEC. Speech-Language and educational consequences of unilateral hearing loss in children. Archives of otolaryngol ogy head & neck surgery. 2004; 130, 524-30.

ABSTRACT

BACKGROUND: In the past, unilateral hearing loss (UHL) in children was thought to have little consequence because speech and language presumably developed appropriately with one normal-hearing ear. Some studies from the 1980s and 1990s have suggested that a significantly increased proportion of children with UHL may have educational and/or behavioral problems, compared with their normal-hearing peers. Limited data exist about the effect of UHL on acquisition of speech and language skills.
OBJECTIVE: To review the current literature about the impact UHL has on the development of speech and language and educational achievement.
DATA SOURCE: MEDLINE search between 1966 and June 1, 2003, using the medical subject heading "hearing loss," combined with the textword "unilateral."
STUDY SELECTION: Studies were limited to those written in English, reporting speech-language and/or educational results in children.
DATA EXTRACTION: Articles were read with attention to study design, population, recruitment of subjects, and outcomes measured.
DATA SYNTHESIS: Problems in school included a 22% to 35% rate of repeating at least one grade, and 12% to 41% receiving additional educational assistance. Speech and language delays have been reported in some but not all studies.
CONCLUSIONS: School-age children with UHL appear to have increased rates of grade failures, need for additional educational assistance, and perceived behavioral issues in the classroom. Speech and language delays may occur in some children with UHL, but it is unclear if children "catch up" as they grow older. Research into this area is necessary to clarify these issues and to determine whether interventions may prevent potential problems.

9. Fellinger J, Holzinger D, Beitel C, Laucht M, Goldberg DP. The impact of language skills on mental health in teenagers with hearing impairment. Acta Psychiatrica Scandinavica. 2009; 120 153-59.

ABSTRACT

OBJECTIVE: The aim of this study was to examine the relationship of language competence level and mental distress in teenagers with hearing impairments.
METHOD: 43 pupils were given a battery of linguistic tests and the Strengths and Difficulties Questionnaire (SDQ), which was also completed by 40 parents. Comparisons were made between the group of 33 children in mainstream education and 10 who were in a segregated school for the deaf.
RESULTS: The children had impaired language skills relative to published norms, especially marked in segregated schools. Parents rated children as having more distress than published norms. Those with superior level of spoken language had fewer peer relationship problems in mainstream education, but significantly more in segregated schools. The reverse was almost significant for those proficient in signed language.
CONCLUSION: Peer relationship problems are associated with the language competence levels in the way that children at school communicate with one another.

10. Kunst SJ, Hol MK, Mylanus EA, Leijendeckers JM, Snik AF, Cremers CW. Subjective benefit after Baha system application in patients with congenital unilateral conductive hearing impairment. Otology & neurotology. 2008 Apr;29(3):353-58.

ABSTRACT

OBJECTIVE: To study whether unilateral Bone-anchored Hearing Aid (BAHA) fitting led to subjective hearing benefit in patients with congenital unilateral conductive hearing impairment.
STUDY DESIGN: Prospective evaluation on 20 patients.
SETTING: Tertiary referral center.
PATIENTS: 10 adults and 10 children with congenital unilateral conductive hearing impairment, with a mean air-bone gap of 50 dB, were included.
METHODS: Subjective bilateral hearing benefit after Baha fitting was measured using 2 disability-specific questionnaires: Chung and Stephens and the Speech, Spatial and Qualities of hearing profile (children's version in the patients aged <18 yr). The Glasgow children's benefit inventory was also used to measure patient's health benefit after Baha fitting.
RESULTS: Chung and Stephens' questionnaire showed an overall preference for the Baha in several specific hearing situations. The Glasgow children's benefit inventory demonstrated an overall mean improvement of +34, which was the most prominent in the learning domain. The 10 adults showed an already good score on the Speech, Spatial and Qualities of hearing scale in the unaided situation.
CONCLUSION: The Baha was well accepted by most of the patients with congenital unilateral conductive hearing impairment.
A preoperative trial of the Baha system with the Baha on a headband is part of the preoperative procedure. In children with unilateral conductive hearing loss, with regard to possible childs' development and communication difficulties, intervention with Baha can be considered as an option.

11. Nicholson N, Christensen L, Dornhoffer J, Martin P, Smith-Olinde L. Verification of Speech Spectrum Audibility for Pediatric Baha Softband Users with Craniofacial Anomalies. Cleft Palate Craniofacial Journal. 2010 Feb;22.

ABSTRACT

Objective: The purpose of this study was (1) to determine benefit of the Baha Softband coupled to the Softband for infants and children with bilateral conductive hearing loss; and (2) to verify audibility of the speech spectrum for octave frequencies 500 through 4000 Hz. Design: The research design for this retrospective chart study is pretest-posttest repeated measures. Setting: The study was conducted in the Department of Audiology and Speech Pathology, Arkansas Children's Hospital. Participants: Twenty-five children aged 6 months to 18 years with craniofacial disorders and bilateral conductive hearing loss participated in the study. Participants were consistent, full-time unilateral Baha users with the Baha Compact bone-conduction amplifier coupled to the head via the Softband. Interventions: The intervention was the Baha device coupled to the head via the Softband as a prerequisite to surgical implantation. Main Outcome Measure(s): The primary study outcome measures used aided and unaided soundfield audiometric thresholds to calculate functional gain. Audibility of the speech spectrum was verified by comparison with target aided thresholds. Results: Results revealed an improvement in soundfield thresholds with Baha amplification for the four octave frequencies. Means, standard deviations, and confidence intervals for aided and unaided thresholds are reported. Percentages of thresholds meeting target levels were significant at all frequencies,

exceeding the 80% criterion. Conclusions: Benefit of the Baha in providing audibility of the speech spectrum for infants and children with bilateral congenital conductive hearing loss has been demonstrated, offering important and timely data supporting third-party reimbursement.

12. Verhagen CV, Hol MK, Coppens-Schellekens W, Snik AF, Cremers CW. The Baha Softband A new treatment for young children with bilateral congenital aural atresia. International Journal of Pediatric Otorhinolaryngology. 2008;72, 1455—1459.

ABSTRACT

The Baha (bone-anchored hearing aid) Softband appears to be an effective mean of hearing rehabilitation for children with a congenital bilateral aural atresia who are too young for the amplification of a Baha on an implant. The aided hearing threshold with a Baha Softband is almost equal to that achieved with a conventional bone conductor. The speech development of the children studied with a Baha Softband is on a par with peers with good hearing.

13. Dun CA, de Wolf MJ, Mylanus EA, Snik AF, Hol MK, Cremers CW. Bilateral bone-anchored hearing aid application in children: the Nijmegen experience from 1996 to 2008. Otology & neurotology. 2010 Jun;31(4):615-23.

ABSTRACT

OBJECTIVE: This study presents clinical data and quality of life questionnaire outcomes in children and young adults with bilateral Bone-Anchored Hearing Aids (Bahas).
STUDY DESIGN: Retrospective review.
SETTING: Tertiary care referral center.
PATIENTS: Eligible study subjects comprised 27 patients with bilateral conductive hearing loss fitted with bilateral Bahas in childhood or as young adults at the Radboud University Nijmegen Medical Centre between June 1996 and October 2008.
METHODS: Questionnaires comprised the "Daily use of bilateral Bahas" questionnaire, the Glasgow Children's Benefit Inventory, and the Speech Spatial and Qualities of Hearing scale modified for children.
RESULTS: A total of 23 children were selected to fill out the postal questionnaires; 21 (91%) of them responded. In 90%, both Bahas were being used 7 days a week. One child was using 1 Baha but not the other, and one child was only using both Bahas at school. Nine children reported that they switched off both Bahas when the background became too noisy. Bilateral Bahas provided better hearing quality according to 70%. The Glasgow Children's Benefit Inventory demonstrated subjective overall benefit of +38 (n = 20). The spatial domain of the Speech, Spatial and Qualities of Hearing scale showed a trend toward better spatial hearing with decreasing age at bilateral application.
CONCLUSION: Bilateral percutaneous Bahas can be applied simultaneously from the age of 4 years, after a successful trial period with the Baha Softband or other equivalent hearing aids. Bilateral Bahas showed clear benefit in the vast majority. Outcomes of bilateral audiologic testing are needed in the near future.

14. Saliba I, Woods O, Caron C. Baha results in children at one year follow-up: a prospective longitudinal study. International journal of pediatric otorhinolaryngology. 2010 Sep;74(9):1058-62.

ABSTRACT

OBJECTIVE: To evaluate audiometric and clinical results of children fitted with a bone-anchored hearing aid with specific emphasis on speech discrimination in different sound environments after one year of use.
METHODS: We performed a prospective longitudinal study. Seventeen patients between the ages of 5 and 18 years old were included. All patients underwent a complete tonal and vocal evaluation at four pre-determined intervals between the pre-operative period and one-year of bone-anchored hearing aid (Baha) use. Basic pure-tone average and speech reception threshold were measured in different sound environments. Speech discrimination improvement was tested with the voice originating from the side of the Baha-fitted ear and with the voice originating from a source directly in front of the patient. These measures were repeated with confounding noise facing the patient then from the side of the affected ear. All tonal and vocal evaluations were performed pre-operatively, the day of processor insertion, 6 months and 12 months after processor insertion. A variance analysis was performed to compare differences in hearing gain with Baha over time.
RESULTS: Hearing gain with Baha was clinically and statistically significant at all intervals. Conventional tonal evaluation revealed significantly improved hearing gain after Baha insertion compared with pre-operative testing with Baha (26.3 dB vs. 17.3 dB), and this improvement was maintained at one year (27.9 dB). Speech discrimination gain at one year was better than immediately post-insertion (21.9% vs. 11.7%). Maximal gain with Baha was found with the voice originating from the side of the affected ear and with confounding noise facing the patient (27.1% at one year), whereas the least gain was found in a silent room with the voice coming from straight ahead (11.9% at one year).
CONCLUSIONS: Pure-tone average gain at one year post-insertion was similar to immediate post-insertion gain. Baha aids speech discrimination most when the voice originates from the side of the affected ear with confounding noise facing the patient. Speech discrimination gain improves with time, suggesting an underlying learning process. The best Baha gain in speech discrimination occurred with background noise.

15. van der Pouw KT, Snik AF, Cremers CW. Audiometric results of bilateral bone-anchored hearing aid application in patients with bilateral congenital aural atresia. Laryngoscope. 1998 Apr;108(4 Pt 1):548-53.

ABSTRACT

The effect of bilateral application of bone-anchored hearing aids (Bahas) was examined in terms of directional hearing and speech recognition in quiet and in noise in four patients with bilateral congenital atresia who, out of pure necessity, had been using a unilateral bone-conduction hearing aid since early life. This study comprised a prospective clinical evaluation in a single subject design; four patients with bilateral congenital atresia originating from the Nijmegen Baha series participated. Three patients had Treacher Collins syndrome. All four patients had conductive, most probably, symmetrical, hearing loss. Recently these patients had applied for a second Baha and were subsequently fitted bilaterally. With two Bahas, all four patients showed significant improvement in sound localization. Also, speech perception in quiet showed significant improvement with bilateral application, and a significant improvement was found in speech perception in noise in three patients. These results suggest that patients with congenital conductive, symmetrical hearing loss will benefit from bilateral Baha.

16. McLarnon CM, Davison T, Johnson IJ. Bone-anchored hearing aid: comparison of benefit by patient subgroups. Laryngoscope. 2004 May;114(5):942-4.

ABSTRACT

OBJECTIVES/HYPOTHESIS: The osseointegrated bone-anchored hearing aid, using the Brånemark system, is well established and has proven benefit. The aim was to study quality of life benefits within patient subgroups using the validated Glasgow Benefit Inventory (GBI).
STUDY DESIGN: Retrospective questionnaire study.
METHODS: Ninety-four consecutive patients were enrolled into the study. Mean patient age was 49 years, with a female-to-male ratio of 1.1:1. Patient subgroups were discharging mastoid cavities, chronic active otitis media, congenital ear problems, otosclerosis, and acoustic neuroma and other unilateral hearing losses.
RESULTS: The response rate was 73%. The score for total benefit of bone-anchored hearing aid fitting for the entire group was +33.3 (95% confidence interval [CI], 25-42). Glasgow Benefit Inventory scores for each subgroup were all greater than +20. The congenital atresia group scored highest with +45 (95% CI, 28-61). Variation in benefit across the subgroups has been demonstrated. Fitting of Baha following acoustic neuroma surgery was shown to be of benefit with a score of +22.2. General benefits scored highest in all subgroups compared with physical and social benefits.
CONCLUSION: The study demonstrated the differences in benefit within patient subgroups. Its results can be used to give patients a predictive value at the time of preoperative counseling. The study identified congenital ear disorders as the group likely to obtain maximal benefit. Notably, for the first time, the study demonstrated the documented benefit of restoring stereo hearing to patients who have acquired unilateral hearing loss following acoustic neuroma surgery using a Baha.

17. Evans AK, Kazahaya K. Canal atresia: "Surgery or implantable hearing devices? The experts question is revisited". Journal of Pediatric Otorhinolaryngology. 2007;71, 367-374.

ABSTRACT

Objectives: (1) Evaluate hearing results in patients managed with external auditory canal reconstruction; (2) compare results to the expectations from treatment with surgical; placement of an osteo-integrated bone-conduction device (Baha system);
(3) assess complications of both interventions; (4) evaluate the medical cost-effectiveness of each avenue of management at 2005 rates for billings based upon relative value units (RVUs). Methods: (1) Retrospective chart review for 36 ears in 29 pediatric patients who underwent surgical canal reconstruction at a tertiary-care pediatric hospital in a major urban center with assessment of management techniques and surgical and audiologic outcomes. (2) Retrospective chart review of six pediatric patients who underwent Baha placement in a major urban center with assessment of management techniques and surgical and audiologic outcomes. (3) Cost reassessment at 2005 billings rates based upon RVUs for canal reconstruction versus BAHA system and comparative analysis. Results: (1) The average post-operative hearing loss in the right ear was 34.3 dB left ear was 31.6 dB. The average gain per ear was 17.3 dB. (2) Twenty-seven (93%) of EAC reconstruction patients required some form of amplification post-operatively. (3) Data available for three of the BAHA patients reflected the predicted average gain in dB (predicted 34.3 dB, observed 31.8 dB). (4) Early complications of canal reconstruction included removal of the packing by the patient, post-operative bleeding and postoperative hematoma. (5) Late complications included recurrent canal stenosis, recurrent otitis externa, canal prolapse and canal cholesteatoma. (6) Uncomplicated external auditory canal reconstruction cost $51505.98 or $2909.94/dB of hearing gain based upon billings fro RVUs. (7) Uncomplicated two-staged Baha system placement cost $42448.85 or $1237.57/dB of hearing gain based upon billings for RVUs. (8) Uncomplicated single-staged Baha system placement cost $28341.00 or $826.27/dB of hearing gain based upon billings for RVUs. Conclusions: Study results indicate that even with significant investments in EAC reconstruction, most patients still required some form of amplification. There are also significant risks of early and late complications from the reconstructive procedure. Studies indicate and our results support that the osteo-integrated bone-conduction device (Baha system) can achieve truly acceptable hearing (_15 dB) in school-aged children with normal bone curves, and it can match the bone-curves for children with sensorineural hearing loss. The two-staged Baha system placement may be provided at almost one-third the cost to the medical system, on a decibel-for-decibel basis. The single-stage Baha system placement yields even greater cost savings at just over one quarter of the cost of surgical EAC reconstruction on a decibel-for-decibel basis. Additionally, there are fewer complications and fewer follow-up visits for the care of the implant system. This is an added value to the parents and patient for the savings in opportunity costs related to the lost time at work and school for office visits and perioperative care. Overall, it appears that osteo-integrated bone-conduction devices may provide a higher quality of outcome for patients while resulting in significant economic savings.

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