

Critical Review: The Effect of a Cochlear Implant compared to Traditional Amplification on Speech and Language Skills for persons with Large Vestibular Aqueduct Syndrome

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This critical review examines the speech and language outcomes of children and adults who, once identified with a Large Vestibular Aqueduct Syndrome (LVAS), become successful users of a cochlear implant. Overall, research supports the current thoughts that a cochlear implant will lead to improved speech and language skills. However, there is a disparity in the results as it has been found that a cochlear implant will benefit some people and not others. At present, a concrete statement regarding the benefits of a cochlear implant with LVAS patients and improvement in speech and language abilities can not be made, due to the limited research directed at this question and the rarity of known affected individuals with LVAS.

Introduction

In recent years, concerns in the audiological field have been raised about the appropriate age and candidacy criterion for implanting a patient with a cochlear device. In the process of evaluating a candidate for a cochlear implant, other factors begin to surface. One apparent factor is the stability of hearing levels. Audiologists can be faced with a client who exhibits a rare hearing loss known as the Large Vestibular Aqueduct Syndrome (LVAS). This was first identified by Valvasorri and Clemis in 1978 as a hearing loss that fluctuates and progressively worsens overtime. It can be a sensorineural, conductive or mixed hearing loss in nature.

Many thoughts exist in today's literature about what LVAS is and how it is caused. Although there is at present no consistent definition of LVAS, one current thinking supports the notion that LVAS can be a result of minor head injuries. However the prevailing research suggest that LVAS occurs when the bony canal connection between the endolymphatic sac and the vestibule in the inner ear stops development by the fifth gestation week. The vestibular aqueduct remains large and never develops normally into a narrow and long structure. The aqueduct is said to be large when it is equal to, or greater than, 2 mm wide. It more frequently occurs bilaterally and affects more males than females.

LVAS is a non-syndromal hearing loss and can occur in isolation or in combination with other symptoms such as Pendred or branchio-oto-renal syndrome. At present, the majority of hearing treatment options for LVAS is via a hearing instrument. Due to the nature of the hearing loss, a cochlear implant becomes warranted once the hearing loss reaches to an unaidable listening range whereby the traditional amplification has been proven to be

ineffective. In most cases, persons with LVAS have acquired a hearing loss over a period of time. However, within this period of time, speech and language skills have already been acquired. Consequently, a cochlear implant can be used to enhance their post lingual speech and language skills.

Objectives

The primary objective of this paper is to critically evaluate the existing literature regarding the benefits of a cochlear implant versus traditional amplification in patients with LVAS and the outcomes of speech and language development skills in this sub-population.

Methods

Search Strategy

Computerized databases included PUBMED and OVID using the following search strategy: (Large Vestibular Aqueduct Syndrome) AND (cochlear implant) OR (Large Vestibular Aqueduct Syndrome) AND (outcome measures) AND (cochlear implant)

Selection Criteria

Studies selected for inclusion in this critical review paper were required to investigate the outcomes in speech and language development in LVAS patients using a cochlear implant device. There were no limits placed on age or demographics. However, this review looked at studies where LVAS occurs in isolation and had no other confounding factors such as in Pendred syndrome.

Many of the participants in this review were children with the exception of one study that looked at adults. This review is supported by a few articles that evaluated the outcomes of speech and language skills of cochlear implant users who were diagnosed

with LVAS. All of the participants used traditional amplification and as their hearing levels worsened, the hearing aids became ineffective and a cochlear implant was seen as a more viable treatment option.

Data Collection

Results in the literature search yielded five articles that were congruent with the selection criteria: case study (1) and retrospective reviews. (4)

Results

Five studies have been conducted to determine the speech and language gains in cochlear implanted children and adults with LVAS.

Au and Gibson (1999) studied 10 children with profound hearing loss at a cochlear implant centre. Over the course of three years, the hearing levels in all of the participants declined to a range whereby a cochlear implant was warranted. The children in this study all showed evidence from radiological assessments that the vestibular aqueduct was wider than 2 mm. The average age of implantation was 6.8 years. All children were unilaterally implanted with a Nucleus Multichannel Cochlear implant with the exception of one child who received a bilateral cochlear implant.

The Bamford-Kowal-Bench (BKB) sentences and the Phonetically Balanced Kindergarten word list (PBK) were open-set words used to assess the child's listening skills in sentences and syllables.

Pre-surgical measures of listening performance revealed poor listening skills. Post surgical measures at 6, 12 and 36 months revealed significant improvements in listening abilities. Overall, the children improved at each assessment and scored from "31% to 79% on BKB tests; from 8% to 43% on average word score and from 38% to 70% on average phoneme score" in six months. (p.186) From six months to three years, there were slight improvements on each test, but none that were significant. Overtime, these children showed significant benefits of a cochlear implant as a viable route to aural rehabilitation and thus as an enhancer of their speech and language skills. Based on parental and teacher's reports, they were able to integrate more with their normal hearing peers and did not require as much external hearing support personnel as required prior to the surgery.

Bent, Chute and Parisier (1999) conducted a study with eight children with a mean age of 8 years old. All of these children had a bilateral sensorineural hearing loss (SNHL) and received limited benefit from amplification. The PBK words and phrases were used, including common phrase sentences. One of the children had "global developmental delays"

(p.1021) and could not produce speech because of presenting cognitive issues. The remaining seven children were reported to have "80% or better on closed set recognition within 12 months on monosyllables and common phrase sentences." (p.1021) Bent et al (1999) concluded that children with LVAS can "recognize open-set words, phonemes and sentences"(p.1022) and can become integrated in the mainstream educational curriculum with the assistance of a cochlear device.

Harker et al (1999) studied five children with an enlarged vestibular aqueduct who had a profound hearing loss between the ages of 4 to 13 years and received a cochlear implant. They were evaluated before cochlear implant surgery with a hearing aid and then after surgery at 3, 6, 12 and 18 months. The PBK test along with common phrases and sentences were used.

There was quite a variation in results among the participants in each of the tests. Four out of the five children showed 80% or better performance on all of the tests and greater achievements were noted the longer the cochlear implant device was worn at 12 and then at 18 months. All of the children were expected to improve at 18 months and beyond. Better scores on sentences versus words in isolation were achieved and this might be because more content information is available in sentences than in isolated words alone.

Temple, Ramsden, Axon and Saeed (1999) diagnosed 15 people out of 350 people who were referred to a cochlear implant centre with LVAS. Seven of these patients with LVAS received a cochlear implant. (5 adults, 2 children) Of the 5 adults, 3 of them performed with 64%, 74% and 100% correct on the BKB tests with open set word recognition in a quiet environment using the cochlear implant device only. (Temple et al, 1999) The other two adults and children were not performing well and the authors noted that more time span was needed to evaluate their performance. Temple et al (1999) did mention that although the results are not very convincing with the children, at the same time, the adults were demonstrating improved speech recognition with the cochlear implant device. A limitation to this study was that the BKB test was not scored prior to the cochlear implant surgery and there is no comparison of performance that can be made at the pre and post surgery times.

Clark and Roeser (2005) conducted a case study report with a female toddler who had fluctuating and progressive SNHL. Her present hearing aids were inadequate and a cochlear implant was recommended and subsequently performed. Two months post implant, reports demonstrate significant gains in four tests that were used to assess speech and

language skills. All of the tests used were to assess the child's awareness to sounds and speech, which were poorly represented prior to the surgery.

The studies described above used a few subjects and this can be justified because of the rarity of this type of hearing loss that occurs in the general population. Random assignment to groups and random sampling was not possible given the above reasons. The participants were seen over a short period of time after surgery and at present, there are no longitudinal studies that evaluate the speech and language skills in patients with LVAS and with a cochlear implant.

Recommendations for Clinical Practice

Although the available literature has provided some evidence of a benefit of a cochlear implant in patients with LVAS and positive outcomes in the acquisition of speech and language skills, there are some limitations to this research. Some studies have shown that a cochlear implant does provide a critical route for speech and language to develop and is a viable option especially in the younger population. However, some of the studies have shown a disparity of results in the outcomes of performing a cochlear implant among subjects. Despite the small sample size that were used in each study and given the rarity of this condition, one can see the benefits of individualized treatment that can be followed in a more manageable case by case basis. Overall, many of the subjects were unsuccessful users of a hearing aid due to the nature of the hearing loss. As a result, their hearing instrument was inadequate to meet their daily listening needs and demands.

Hence, a cochlear implant was seen as an optimal route to aural rehabilitation. Most of the subjects in this review have acquired some speech and language skills prior to the cochlear implant and were not pre-lingually deafened. At present, my clinical recommendation would be that although there are individual differences in outcomes from having a cochlear implant and the research has demonstrated that while it is a beneficial surgery, the outcomes are more positive for some people with LVAS than for others in terms of speech and language gains.

The research articles above do not indicate which patients will benefit from a cochlear implant and which would not even with today's sophisticated radiological screening such as magnetic resonance imaging (MRI). There are too many individual differences to initially account for who will and who will not become a successful cochlear implant user.

Conclusion

Although the research is limited in this area, there is support that a cochlear implant does assist some people with LVAS to acquire and maintain their speech and language skills. The amount of benefit at this point is not clear and more research is needed in this topic before a concise statement can be proclaimed.

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