Critical Review: Speech perception outcomes following cochlear implantation in children with non-syndromic Auditory Neuropathy Spectrum Disorder as compared to children with sensorineural hearing loss?

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This critical review examines the speech perception abilities of cochlear implanted children diagnosed with Auditory Neuropathy Spectrum Disorder (ANSD) as compared to implanted children with sensorineural hearing loss (SNHL). Study designs included: between subjects non-randomized intervention studies and mixed (between & within) non-randomized intervention studies. Overall, research suggests that cochlear implantation provides speech perception benefit in some children with ANSD who have demonstrated a lack of success with traditional amplification. However, a definitive statement regarding the post-implant performance of ANSD children relative to SNHL children cannot be made due to research limitations including

(auditory dys-synchrony) AND (cochlear implant). The search was limited to peer-reviewed articles written in English and involving human participants.

Selection Criteria

Studies selected for inclusion in this critical review were required to investigate the speech perception abilities of implanted ANSD children (< 18 yrs) as compared to implanted SNHL children. The studies were limited to those including children with non-syndromic ANSD without associated medical disorders. No limits were set on the age of implantation or the research methods used.

Data Collection

Results of the literature search yielded six articles that were congruent with the selection criteria above: 3 between-groups and 3 mixed groups non-randomized intervention studies. In accordance with the level of evidence hierarchy for high-quality standards (Cox, 2005), all six studies provided a level 3(-) of evidence.

Results

Between Groups Non-Randomized Intervention Studies

Study 1. Buss et. al. (2002) used a prospective design to compare speech production outcomes in children with ANSD (n=4) and children with SNHL (n=33) following unilateral cochlear implantation. Two of the ANSD participants (S1 and S2) were approximately 2 years old at the time of implant and were matched with a group of SNHL children implanted between 2 and 4 years of age (n=13). The other two participants (S3 and S4) were approximately 5.5 years old at the time of implant and matched with a group of SNHL children implanted between 4 and 6 years of age (n=13).

Speech production was assessed at 1 year post-implantation by a speech language pathologist (SLP) according to nine categories of possible errors using the Paden-Brown test. Neural integrity was evaluated by measuring electrically evoked auditory brainstem responses (EABR).

Individual ANSD participant test scores were compared to the mean score of the matched control group. This comparison revealed that participants S1, S2, and S4 had post-implant scores that fell within or above one standard deviation of the control group mean on all nine test categories. Participant S3 had speech production scores that fell more than one standard deviation below the mean of the control group on two of the nine test categories. This result was associated with S3's continued use of manual communication post-implantation. The EABR measure revealed an identifiable wave V for all four implanted ANSD children. The results were interpreted as evidence that cochlear implantation in ANSD children can produce a synchronized neural response and that this can be used

attribute group differences in speech perception to differences in hearing disorder and site of lesion alone.

Study 3. Leigh et. al. (2009) compared post-operative speech perception in implanted ANSD children (n=7) to previously reported outcomes from implanted SNHL children (n= 102). The ANSD group ranged in age from 3.5 to 8.5 years and varied in duration of implant use from 2 to 4 years. Mean age at implantation ranged from 6 months to 4.5 years. Participant

for in some studies (Rance & Barker, 2008; Peterson et. al., 2003) but not others (Buss et. al., 2002; Gibson & Sanli, 2007; Jeong et. al., 2009; Leigh et. al., 2009). Finally, the studies varied in their tendency to control for differences between participant groups on variables known to affect speech perception test results, such as age at implantation, duration of implant use, age at assessment, type of implant, mode of communication and post-implant auditory training. Two of the reviewed studies made no attempt to match participant groups on any of these variables (Gibson & Sanli, 2007; Leigh et. al., 2009). These differences in research design and methodology limit the validity of comparisons across studies.

Based on the aforementioned study limitations, there is not a strong degree of evidence to support the use of cochlear implants as a treatment option for children with ANSD. In fact, Rance & Barker (2008) found that implanted ANSD children performed worse on post-operative speech perception measures than implanted SNHL children. This study had some advantages over the others as the ANSD participants were sub-divided based on degree of benefit from amplification. This study also provided a better description of participant selection by specifying the criteria used to indicate lack of hearing aid benefit and they accounted for the influence of vocabulary and speech production limitations on test outcomes. However, it is possible that the subject selection criteria introduced experimental bias causing the implanted ANSD participants to perform worse than the SNHL group. Specifically, the higher functioning ANSD subjects, less disordered by their condition, were successful with hearing aids and therefore did not receive a cochlear implant. Conversely, those with more disabling ANSD, who did not benefit from amplification, formed the implanted ANSD group. It is unknown whether the aided ANSD participants would have performed better with cochlear implants and, therefore, whether the results were biased in a negative direction.

In contrast, Gibson & Sanli (2007) found that ANSD children demonstrating post-implant neural synchrony performed significantly better than those with abnormal neural responses and children with SNHL. The findings of this study support the notion that the success of cochlear implantation may depend upon underlying pathology. By failing to separate ANSD participants according to post-implant neural status, the other studies included in this review may have used a group of ANSD children that were heterogeneous in underlying pathology. This may have resulted in a large variance in speech perception outcomes and the failure to find significant differences between groups.

Clinical Recommendations

Regardless of the performance of implanted ANSD children relative to implanted SNHL children, studies employing a pre-and post-implant repeated measures design found significant improvements in speech perception abilities of ANSD children following implantation (Jeong et. al., 2009; Rance & Barker, 2008). This finding, along with the results of the study by Gibson & Sanli(2009), suggest that cochlear implantation may be beneficial for some ANSD children, especially those with a peripheral rather than retrocochlear site of lesion. However, given the available research, it remains difficult to predict the success of this treatment for individual ANSD children. This uncertainty, along with the permanent nature of cochlear implantation, means that a strong recommendation for its use as a standard treatment for all ANSD children cannot be made.

The assumption that ANSD children will not benefit from traditional amplification has resulted in the use of cochlear implantation as the default treatment strategy for this population. However, it has been demonstrated that hearing aids may be a viable option in some cases of ANSD (Rance & Barker, 2008). Therefore, all ANSD children should undergo a rigorous trial period with simultaneous amplification and intensive auditory-oral habilitation similar to that provided for cochlear implant recipients. In addition, clinical practice guidelines outlining the parameters of such hearing aid trial

Gibson, W.P. & Sanli, H. (2007). Auditory Neuropathy: An update: *Ear and Hearing, 28(2), 102S-106S*

Jeong, S.W., Kim, L.S, Kim, B.Y., Bae, W.Y & Kim J.R. (2009). Cochlear Implantation in children with auditory neuropathy: Outcomes and rationale. *Acta Oto-Laryngologica, 127 (10), 36-43*

Leigh J., Rance, G., Shani, D. & Dowell, R.(2009) Cochlear Implant Outcomes for Children with Auditory Neuropathy Spectrum Disorder. *Perspectives on Hearing and Hearing Disorders in Childhood, 19, 75-84.*

Rance, G. & Barker, E.J. (2008) Speech Perception in Children with Auditory Neuropathy/ Dyssynchrony Managed with Either Hearing Aids or Cochlear Implants. *Otology & Neurology, 29, 179-182.*

Peterson, A., Shallop, J., Driscoll, C., Breneman, A., Babb, J. Stoeckel, R. & Fabry, L. (2003). Outcomes of Cochlear Implantation in Children with Auditory Neuropathy. *Journal of the American Academy of Audiology, 14(4), 188-201*.