Cochlear nerve aplasia

The absence of a visible cochlear nerve or cochlear nerve canal on radiologic imaging does not preclude auditory innervation of the cochlea. Cochlear implantation can be a viable option for patients with apparent cochlear nerve aplasia who have undergone appropriate testing. Electronically evoked auditory brainstem response is critical in the evaluation of this patient group [1].

Cochlear nerve aplasia or hypoplasia is found in up to a half of patients with unilateral or bilateral hearing loss. There is an ongoing discussion regarding the indication of cochlear implants (CI) for hearing rehabilitation in cases with radiologically-defined aplasia or hypoplasia of the cochlear nerve in those patients, especially in children. At present there is conflicting evidence whether the audiological outcomes of those children with a CI are comparable to those of children with a CI and a radiologically-normal cochlear nerve.

The primary aim of a study of Ehrmann-Müller et al., from the University of Wuerzburg, Germany, was to assess the audiological abilities before and after cochlear implants (CI) provision in children with cochlear nerve hypoplasia or aplasia. Additionally, they aimed to determine if audiological outcomes differed in children with aplasia from those with hypoplasia. Such data should be helpful in determining if CI provision is appropriate for such children.

This retrospective study presents 7 children who were diagnosed with cochlear nerve aplasia or hypoplasia and received a CI. The pre- and postoperative audiological performance and the hearing and speech development of the children were examined.

4 children were unilateral CI users and 3 were bilateral CI users. Hearing reactions could be detected in all children. Already at first fitting, prompt responses and reactions to songs were observed. The aided thresholds in free field in children with hypoplasia were between 30 and 60 dB. Even in children with aplasia, the results in free field with CI averaged between 30 and 70 dB. Therefore the aided thresholds in children with hypoplasia and in children with aplasia of the CN are similar. It could be demonstrated that hearing reactions improve with the long term use of the implant. Improvement in general development could be observed in all children despite the very heterogeneous conditions and the accompanying handicaps.

The results of this study support the hypothesis that children with radiologically-defined CN hypoplasia or aplasia and detectable responses to electrical or acoustical stimuli can improve their sound detection thresholds and their awareness of sound when provided with a CI [2].

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Warren FM 3rd, Wiggins RH 3rd, Pitt C, Harnsberger HR, Shelton C. Apparent cochlear nerve aplasia: to implant or not to implant? Otol Neurotol. 2010 Sep;31(7):1088-94. doi: 10.1097/MAO.0b013e3181eb3272. PubMed PMID: 20634773.
2)
Ehrmann-Müller D, Kühn H, Matthies C, Hagen R, Shehata-Dieler W. Outcomes after cochlear implant provision in children with cochlear nerve hypoplasia or aplasia. Int J Pediatr Otorhinolaryngol. 2018 Sep;112:132-140. doi: 10.1016/j.ijporl.2018.06.038. Epub 2018 Jun 23. PubMed PMID: 30055722.