Cochlear implant outcome in unusual cases

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Objectives: To describe the clinical experience and characterize the outcomes in special cases of cochlear implantation.

To assess auditory functionality and language development.

Background: Minimal requirement for CI is the presence of an implantable cavity with survived neural elements whose projections connect to the auditory cortex.

Among CI candidates, the presence of inner malformations or associated syndromes has the potential to adversely affect outcomes by reducing both the organization and availability of neural population to stimulate electrically.

Methods: 6 patients: Common cavity (n=2). Incomplete partition type II (n=1). Encephalomalacia (n=1). ANSD (n=2). Before implantation, all children had severe to profound SNHL and failed to make appropriate progress during a hearing aid trial.

Results: All subjects underwent successful implantation.

Cochlear implant mapping: Dynamic process that was challenging with current level setting based on a combination of behavioral and ECAP responses.

Conservative approach: Slow rates of stimulation,current levels set low and increased with time and experience with patient's tolerance. A stable map achieved by 6 months after initial stimulation.

Assessment: The efficacy of CI was evaluated using a stepwise hierarchic assessment for achieving applying IT-MAIS/MAIS score: Effective audibility such that thresholds were significantly improved. Improved speech recognition, which means better postoperative SD% than preoperative ones. Effective speech with improved sentence recognition ability in noisy situations. Effective communication with development of speech production and social communication skills.

Conclusions: Such cases undergoing CI require greater stimuli to obtain an auditory response. Inner ear anomalies as common cavity have poor auditory functionality and the prognosis for developing open set speech perception is poor. Children with IP II and presynaptic ANSD have an excellent prognosis for developing open set speech perception as they have an excellent auditory functionality. Decision making for such children remains difficult when spoken language development is the goal.