Dilemmas in the Treatment of Infants with Congenital Deafness and Questionable Presence of Auditory Nerves

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Background

Rehabilitation of congenital bilateral deafness is usually with cochlear implantation. Absence of an auditory nerve precludes implantation. MRI scans are the most commonly used modality to prove the presence of a nerve. When imaging is inconclusive about the status of the nerve a number of dilemmas arise.

Conclusions

1. When the presence of auditory nerve is questionable but not ruled out by imaging, cochlear implantation is a viable rehabilitation modality. This is in contrast to the previous requirement of demonstrating the presence of an auditory n. on imaging

2. Lack of initial electrical or behavioral responses after implantation does not preclude later favorable outcome. The mechanism of this phenomenon is not well understood.

3. Hypoplastic auditory nerve is a negative predictor for CI outcome.

Pre-Operative Imaging

Congenital deafness is studied with imaging to portray the inner ear and auditory nerves:

CT
- Normal Cochlea
- Abnormal cochlear apexure
- Normal internal auditory canals

MRI
- Normal internal auditory canals (IAC)
- Facial and two vestibular nerves present
- Auditory nerves not depicted clearly

Presentation

Case 1
A 20 months old infant failed new born TEOAE hearing screening. No responses on behavioral auditory testing. On repeated objective testing no TEOAEs, cochlear microphonics or ABRs were recorded.

Case 2
Presented at the age of 4 years, with weighing 10kg and some features of CHARGE syndrome (dysmorphic features, unilateral choanal atresia) but negative for common CHD7 mutation. Repeated ABR: no responses at maximal output, present cochlear microphonics and TEOAE; behavioral responses VDL ~ 70-80dB – auditory neuropathy?

Selected Issues in Decision Making

1. In a patient with congenital deafness and questionable presence of cochlear nerves on imaging
   - Are additional tests (i.e. eABR) mandatory?
   - Which side should be implanted or is bilateral implantation the best choice?

2. If no NRT or behavioral responses are present in the OR and during switch
   Adjustment of stimulation (wider pulse, slower pulse rate etc.) may result in better responses.

3. At what point lack of electrical and behavioral response necessitates other rehabilitation modalities (Auditory brainstem implant? Sign Language? Other?)?

Post-Operative Course

Outcome Case 1
3 years past bilateral simultaneous implantation: identifies closed set words, mimics words with mediocre success, pronounces some words

Outcome Case 2
1 year past left ear implantation: identifies lips, identifies closed set large words, with visual reinforcement, progressive word pronunciation (still limited)

REFERENCES