CI2017 PEDIATRIC
15TH Symposium on Cochlear Implants in Children
JULY 26-29, 2017
SAN FRANCISCO, CA

CI2017 SAN FRANCISCO

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Auditory Brainstem Implant in children with cochlear nerve aplasia/hypoplasia

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Purpose

The aim of this study was to evaluate the audiological results of the ABI in eight children with bilateral aplasia and one child with bilateral hypoplasia of cochlear nerve by direct stimulation of the Cochlear Nuclei.

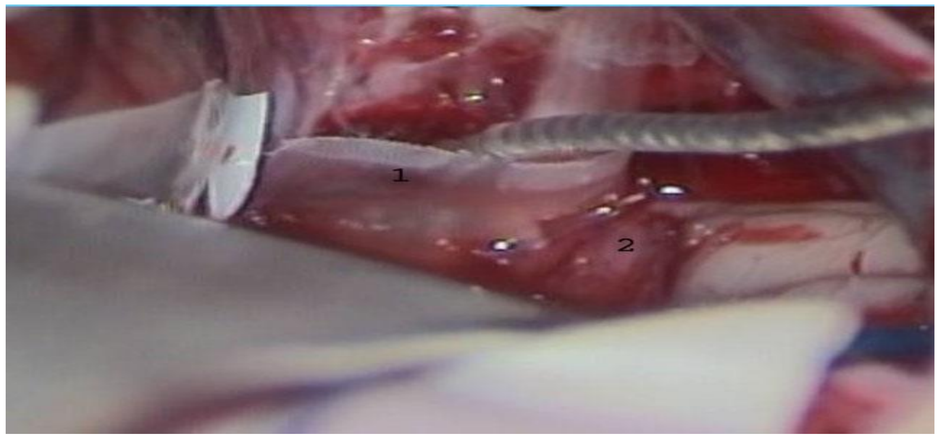
Material and methods

This retrospective study was conducted at the Cochlear Implant Center "Prof. Diamante", Buenos Aires, Argentina.

Children underwent otological, radiological, audiological, neurological, psychological evaluations pre-ABI. In eight children the MRI showed bilateral cochlear nerve aplasia and one child had bilateral hypoplasia of the cochlear nerve. In this child it was performed CI in the better ear without audiological results, so it was made an ABI in the contralateral side.

A Nucleus 24 ABI and Nucleus ABI 541 (Cohlear Ltd) were used in all children with the retrosigmoid approach.

The electrode correct positioning was monitored through the Electrical Auditory Brainstem Recording (eABR).

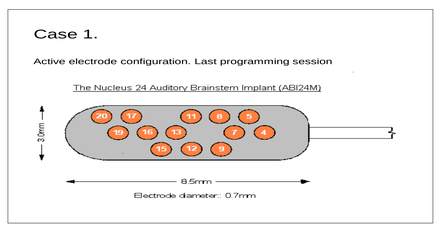


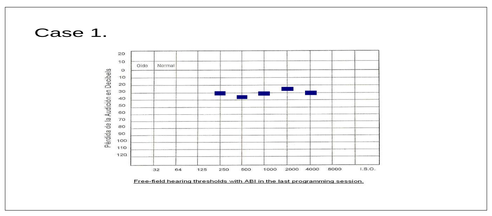
ABI was activated in general, 40 to 60 days after surgery in all patients. Because of the possible risks involved in stimulating brainstem structures, the tune up was done with electrocardiographic monitoring and medical assistance.
The main parameters for electrode activation were Speak strategy , variable stimulation modes (MP1+2 and MP2) and pulse width (100 us, 150us and 200 us).
Mapping was obtained via behavioral observation and play conditioned responses. Initial tune up and control was done during the following days.
The audiological outcomes were evaluated using the CID Speech Perception Categories (Geers, 1994) through tests of the Latin American Protocol, Free Field with ABI, it-MAIS and MAIS scales.

Results

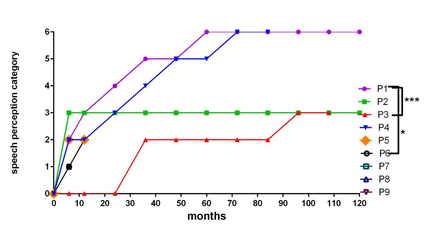
Auditory sensations with the ABI were produced in all patients They used the ABI in a permanent way and showed positive and variable hearing evolution.

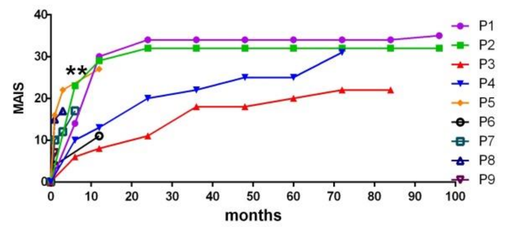
They showed improvement in speech perception (from detection to identification of words from suprasegmental to segmental features) 2/9 children were able to recognize words in Open Set.
All patients use the ABI in a permanent way.
Additional handicaps are higher in this population (6/9)
One child with CI+ABI is performing better with CI AFTER ABI.





Case 1. Active Electrodes and Free Field with ABI
Free Field was around 25-45 dB (freq from 250 Hz to 4000Hz) in all children.

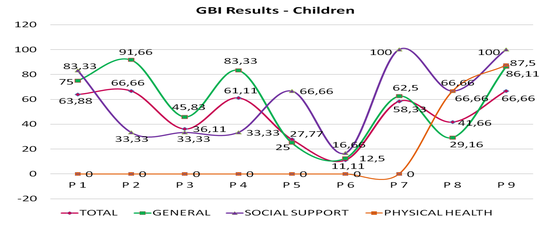




Tukey's multiple comparisons test	Mean Diff.	95% CI of diff.	Significant?	Summary		
P1 vs. P2	1.833	-0.1573 to 3.824	No	ns		A-B
P1 vs. P3	3.129	1.093 to 5.164	Yes	***		A-C
P1 vs. P4	0.9167	-1.233 to 3.067	No	ns		A-D
P1 vs. P5	3.250	0.1026 to 6.397	Yes	*		A-E
P1 vs. P6	3.583	0.4359 to 6.731	Yes	*		A-F
P1 vs. P7	3.250	0.1026 to 6.397	Yes	*		A-G
P1 vs. P8	4.083	0.3593 to 7.807	Yes	*		A-H

Pre-ABI VS 6m	
Paired t test	
P value	0.0086
P value summary	**
Significantly different? (P < 0.05)	Yes

Right) IT-MAIS and MAIS Scales
Left) Categories of Speech Perception (Geers,1994). Evolution



Positive changes in Quality of Life were observed. GBI Test

Conclusions

Structures of the inner ear should be thoroughly investigated prior to ABI to make sure no alternative option is possible.
MRI evaluation, subjective and objective assesment is necessary in congenital deaf children who do not have benefits with hearing aids, in order to determine the ABI indication.
No surgical or postoperative complications were observed.
Surgery and Programming are more complex and longer compared to CI.
Results are poorer and slower when compared to CI children with normal cochlear nerve. Surgery, programming, auditory and language habilitation, other handicaps, parental involvement, age at implant are variables that affect results.This study suggests that ABI could be an effective way of restoring hearing in children with cochlear nerve aplasia/hypoplasia. Our findings are in agreement with previous studies (Colletti et al, 2000, 2001, 2005, 2007; Sennaroglu et al.2009, 2011; Eisenburg et al, 2008, 2015; Choi et al. 2011; Shannon, 2011, Yucel et al.,2015)