Auditory, Speech and Language Outcomes Post Auditory Brain Stem Implant in a Child with Down’s Syndrome

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No relationships to disclose
Outcomes following ABI

- Emerging reports of progress made in auditory function following an ABI.

- There are little to no data on speech, language, cognitive and academic outcomes in pediatric users of an ABI.

- This case study summarizes auditory, speech and language growth in a child following an ABI.
Participant

- Participant is a 5-year-old child with cochlear nerve deficiency (CND) and Down’s syndrome

- She received a cochlear implant at 1 year of age and then an ABI at the age of 3 years, 4 months
Did not pass new born hearing screening
Born at 38-weeks gestation
Stayed in neonatal intensive care due to Patent Ductus Arteriosus (PDA) repair
Experienced feeding difficulties
Developmental milestones were reportedly delayed
MRI revealed absent left and right cochlear nerve

Click ABR produced no response, suggesting a profound hearing loss

Tone burst testing in the left ear showed mod-severe hearing loss in 500-1000 Hz range and severe-profound hearing loss in the 2000-4000 Hz range

P1 auditory evoked potential testing showed no response in the right ear and delayed latency for the left ear
Participant – Device history

- Hearing aids - 8/2009 @ 6mos of age
- Left ear 4/2010 – CI @ 1 yr 2 mos
- Left ear 6/2012 – CI explanted and ABI implanted in Italy @ the age of ~3.5 yrs
- Med-El Concerto -Opus-2 Processor
- ABI activation 07/2012 in Birmingham, AL
Participant: Training

- Participant received auditory training without visual cues for the first year of ABI use at one facility in addition to total communication pre-school.
- No consistent response to sound was seen for the following year.
- Reprogrammed at one year post activation.
- Parents reported child started showing responses to environmental sounds after reprogramming.
During the next 8 months parents and child participated in a multisensory adaptation of Auditory-Verbal therapy at another facility and administered a home program with emphasis on listening and speaking.

During this time, auditory and speech/language development emerged.
Methods

Assessed auditory, language and speech skills using the following:

- Therapy session / progress notes
- Parent reports / instruments
- Reynell Developmental Language scales (RDLS)
- Cottage Acquisition Scales for Listening, Language and Speech (CASLLS)
  - 1-year post activation of the ABI (Time A)
  - 1-year, 8-months post activation (Time B)
- In addition, audio recordings were collected at Time B
Auditory skills

- The participant exhibited auditory awareness of and conditioned response to the Ling 6 sounds presented via live voice.
- Occasionally responded to:
  - name
  - phone ringing
  - door knock
Results – Time A - Baseline

Language skills

RDLS scores revealed:

• Standard score of <63
• Percentile rank of <1 at both time points
Results – Time A - Baseline

Speech production skills

- No spontaneous words
- Spontaneous vocalization to indicate wants
- Approximation of some pitch and durational changes
Auditory skills

- Attempts imitation of all of the 6 Ling sounds delivered through auditory modality alone
- Identifies at least 5 animal sounds without visual cues.
- With auditory cues only, is able to imitate one of the Ling sounds /a/
- Discriminates between vowels: differentiated production of fricatives by approximating /s/ and /sh/
- Follows at least 3 simple directions with speech reading (no signs)
- MAIS SCORE: 23/40
**Language skills**

- RDLS scores unchanged from Time A
  - Standard score of <63
  - Percentile rank of <1
Speech production skills

Participant produces
- emerging word shapes
- variety of vowels
- variety of consonants
- few diphthongs
Speech production skills (cont’d)

Session notes and audio recordings indicated vocalizations consisting of

- variegated babble
- word production
- emerging melodic patterns in spontaneous utterances
Speech production skills (cont’d)

- Imitation of syllable shapes:
  - VC
  - CVCV
  - C1VC2
  - V1CV2
- Vowel and diphthong inventory: i, u, U, ae, au, a o
- Consonants:
  - p, b, m, w, n, h, l
  - emerging /d/, /s/, /sh/
- Vocal turn-taking
Speech production skills (cont’d)

- Spontaneous vocalizations: [badanda] [nda nda] [buabua] [ba ba ba] [ ao ao ao] – variegated babbling
- Spontaneous naming on body parts ma (mouth)
- wa (baby crying) , papa, moo, a-a-a (horse), apple, ba [bye]
- Emerging melody patterns in spontaneous utterances
- MUSS SCORE: 18/40
### Audio samples of speech

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<thead>
<tr>
<th>Syllable shapes</th>
<th>Babbling</th>
<th>Vowels</th>
<th>Consonants</th>
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At the listening age of ~1.5 years

- Progress was seen in auditory skills, speech production skills and vocabulary.

- Preliminary results support the findings of Colletti and Zoccante (2008) that ABI facilitates communication growth in children with associated cognitive deficits.
We want to thank the patient and family for their support of this case report