Case Study

THE IMPORTANCE OF ACCURATE BEHAVIOURAL TESTING IN INFANT HEARING AID FITTINGS

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Asymmetrical Drop in Hearing

- Infant identified from universal newborn hearing screening programme
- Bilateral refer on AABR with 35 dBnHL click stimulus
- Family history of hearing loss
  - Maternal grandfather has loss from early childhood, now has a CI
  - However father convinced hearing loss was due to screening not being done properly
Initial Diagnostic Testing

• 4 ABR appointments required as baby restless, light sleeper

• Type A tympanograms (1000 Hz probe tone)

• Absent ipsilateral acoustic reflexes (1000 Hz probe tone) using broad band noise stimulus

• Absent DPOAEs bilaterally
Toneburst ABR results

Left 2kHz
70 dBnHL

Right 2kHz
70 dBnHL
4kHz toneburst ABR results

Right

Left
Right click (80 dBnHL) ABR

Subtracted

Rarefaction and Condensation overlaid

Alternating

Left ear results similar, no sign of ANSD (no cochlear microphonic)
Absent DPOAEs in both ears

Summary of results from diagnostic audiology appointments (NR=no response)

<table>
<thead>
<tr>
<th></th>
<th>500 Hz</th>
<th>1000 Hz</th>
<th>2000 Hz</th>
<th>4000 Hz</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Toneburst ABR</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left AC</td>
<td>65</td>
<td>65</td>
<td>70</td>
<td>70</td>
</tr>
<tr>
<td>Right AC</td>
<td>65</td>
<td>65</td>
<td>70</td>
<td>85</td>
</tr>
<tr>
<td>BC Left &amp; Right</td>
<td>NR 60</td>
<td>NR 60</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Click ABR</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left &amp; Right AC</td>
<td>Small wave V present; no evidence of ANSD</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td><strong>Tympanometry</strong></td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td>Type A tympanograms</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent ipsilateral acoustic reflexes</td>
<td></td>
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<td></td>
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<tr>
<td><strong>DPOAEs</strong></td>
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<td></td>
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<tr>
<td>Absent left and right whenever tested</td>
<td></td>
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Hearing Aid Fitting

- Fitting at approximately 4-5 months of age

- Parents wanted 2nd opinion, took child to United States to the House Ear Institute (who agreed with our results)

- Bilateral Phonak Bolero Q70 M13
Example of real ear measures

Measured RECDs

Standard speech input at soft, average and loud and MPO check with 90 dB signal

DSL 5.0 fitting method
Ongoing testing and monitoring

• Child receiving regular Auditory Verbal Therapy (AVT) sessions, parents were reasonably happy with his progress

• Very good hearing aid use, highly motivated parents

• Behavioural testing using VRA (Visual Reinforcement Audiometry) began at about 6 months of age in order to check progress with amplification and monitor behavioural thresholds
VRA at 8 months

“not conditioned, not responsive even to live voice”
VRA at 9 months

Still not ready for any frequency specific testing

Monitored live voice testing at 50dBHL?

Not very consistent with ABR thresholds
VRA at 10 months

Parents and audiologists getting concerned over lack of responsiveness to sounds when being tested

Everyone getting a bit stressed
VRA at 11 months

“Had to condition, then was interested, gave some searches, then gave up”

“child became upset, dad concluded appointment”

Discussions begun about repeating ABR
VRA at 15 months

4 appointments later

Finally started doing VRA

Good reliability, but thought they might be supra-threshold
VRA at 16 months

Very reliable
Happy to tolerate inserts
Now unexpected asymmetry apparent

Everyone including parents want to have repeat ABR to check on thresholds
Cortical Evoked Potential Testing

• Child was non-compliant and session abandoned, parents very keen to repeat ABR
GA (General anaesthetic) ABR

- **Left ear**: 4kHz 80 dBNHL (response at 70 dBNHL at initial test)

- **Right ear**: NR at 105 dBNHL (response at 85 dBNHL previously)
GA ABR

Right Click 95dBnHL

Subtracted
Rarefaction
Condensation
Alternating
Right NR

Left Click 95 dBnHL

Left Small
wave V at 95 dBnHL
No CM present
Left 4kHz
80 dBnHL
(previous test threshold=70)

Left 2kHz
85 dBnHL
(previous test threshold=70)
Real Ear Measures

Re-fitted for right profound loss

Naida IX UP
Measured RECDs
Confirmed Genetic Cause of Hearing Loss

<table>
<thead>
<tr>
<th>Sample no:</th>
<th>Gene</th>
<th>Exon</th>
<th>Nucleotide change</th>
<th>Amino acid change</th>
<th>Genotyping results</th>
<th>Zygosity</th>
<th>Type of variation</th>
<th>Verified by sequencing</th>
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<tbody>
<tr>
<td>HF0890</td>
<td>GJB2 (Connexin2 6)</td>
<td>ex2</td>
<td>c.283G&gt;A</td>
<td>p.Val95Met</td>
<td>AG/CT</td>
<td>HET</td>
<td>Mut</td>
<td>+</td>
</tr>
<tr>
<td>HF0890</td>
<td>GJB2 (Connexin2 6)</td>
<td>ex2</td>
<td>c.35delG</td>
<td>p.Gly12fs</td>
<td>GT/AC</td>
<td>HET</td>
<td>Mut</td>
<td>+</td>
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Current progress

• Fitted with a cochlear implant in his right ear and continues to wear his hearing aid in his left ear

• Doing very well and age appropriate for speech and language development

• Very robust cortical evoked potentials present