

Case Study



THE IMPORTANCE OF ACCURATE BEHAVIOURAL TESTING IN INFANT HEARING AID FITTINGS

Andrea Kelly, PhD, MNZAS
Auckland District Health Board
Suzanne Purdy, PhD, MNZAS
University of Auckland

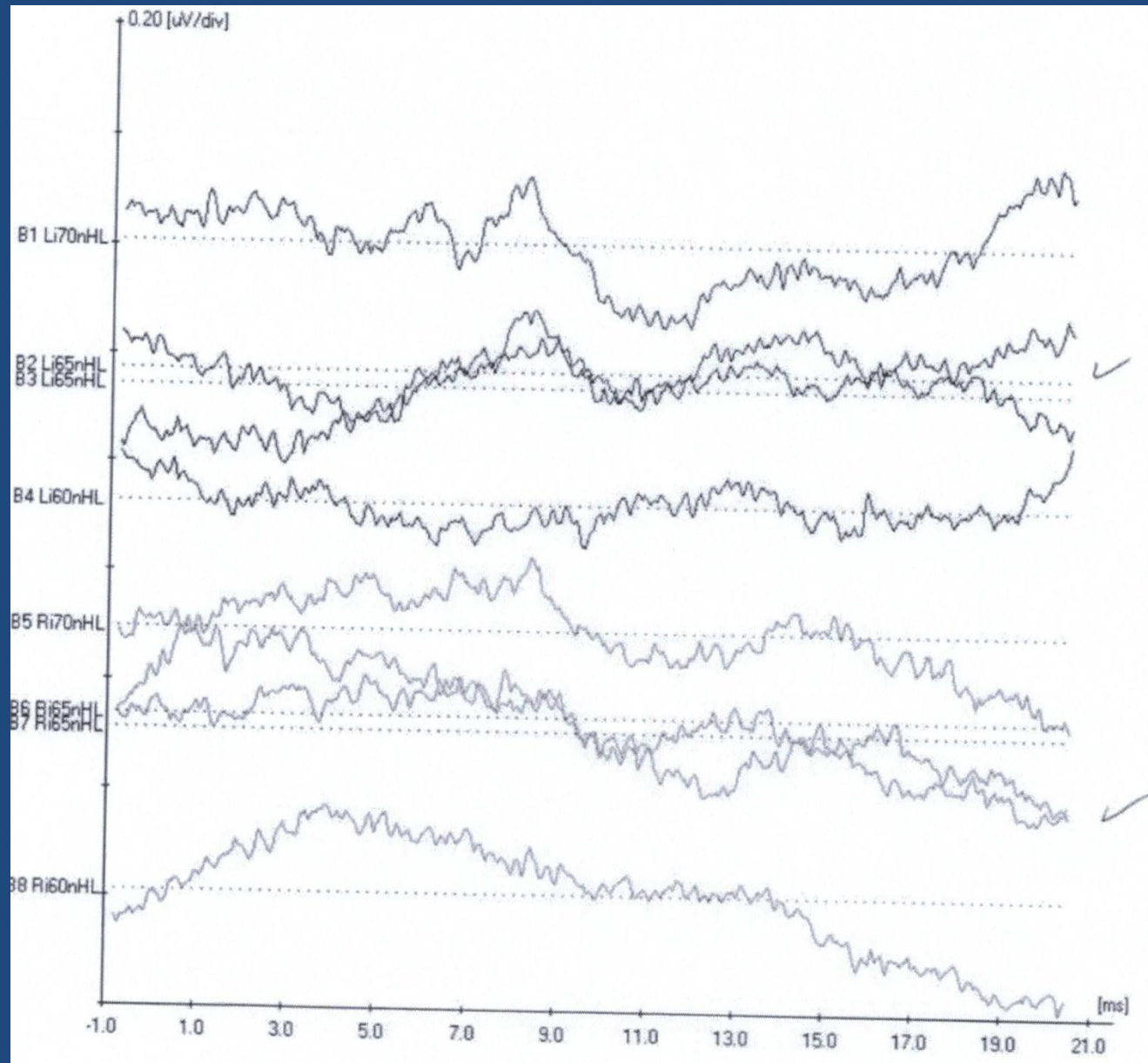
Asymmetrical Drop in Hearing

- 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

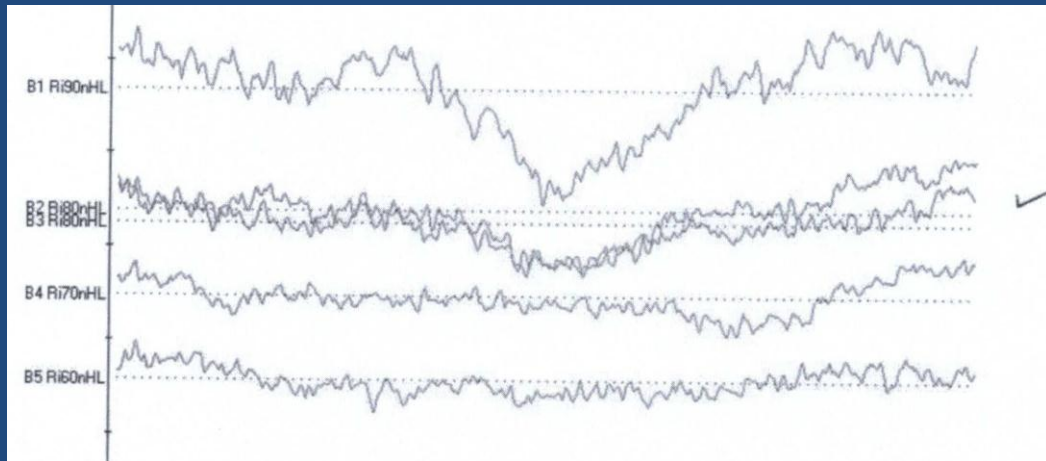


Left 2kHz
70 dBnHL

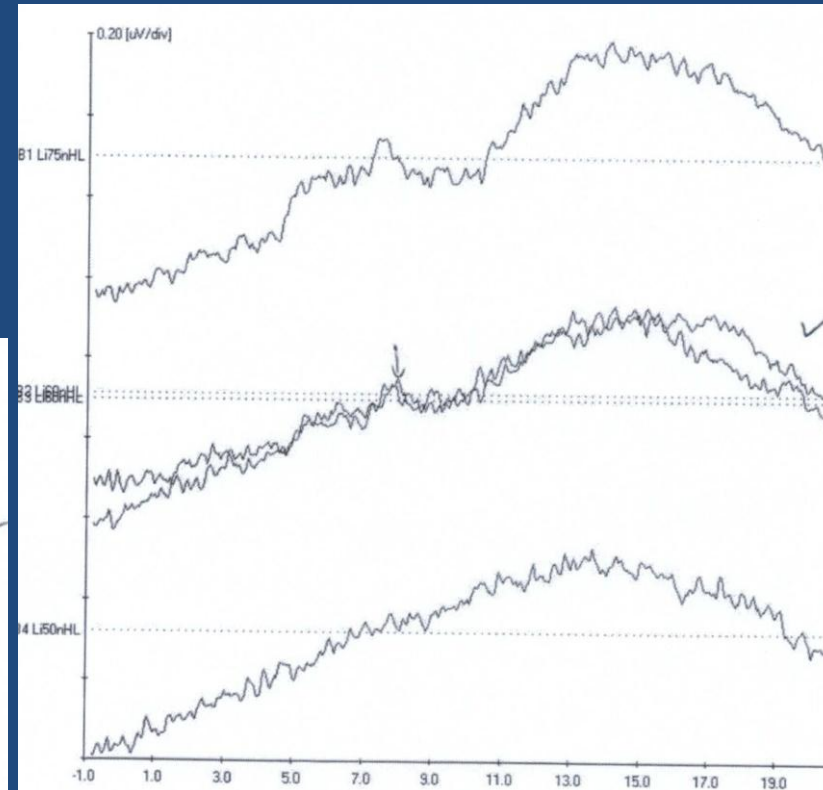
Right 2kHz
70 dBnHL

4kHz toneburst ABR results

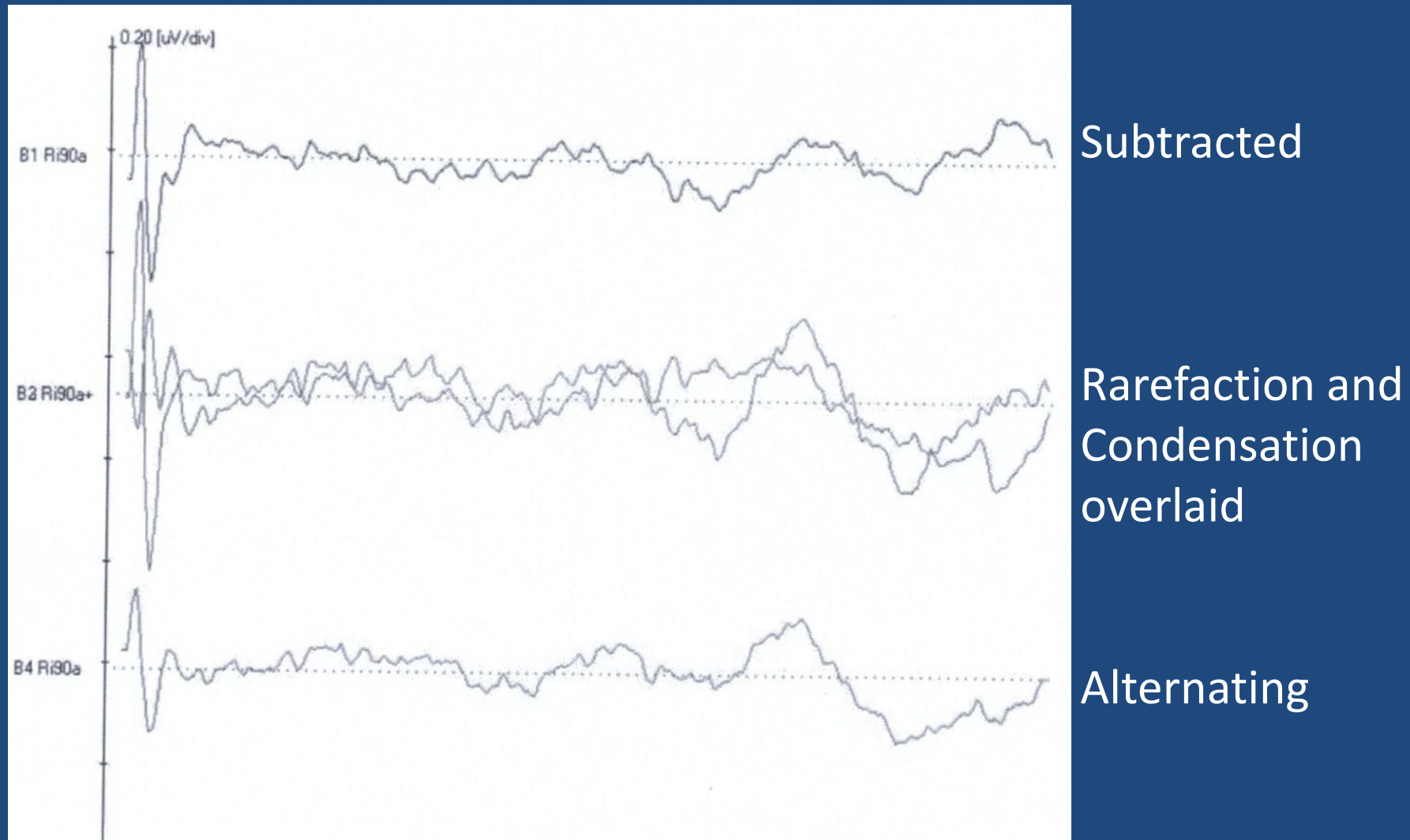
Right



Left

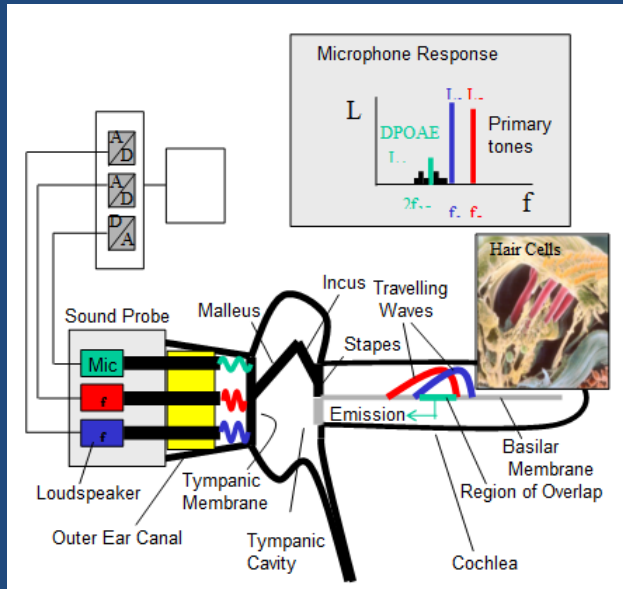


Right click (80 dBnHL) ABR

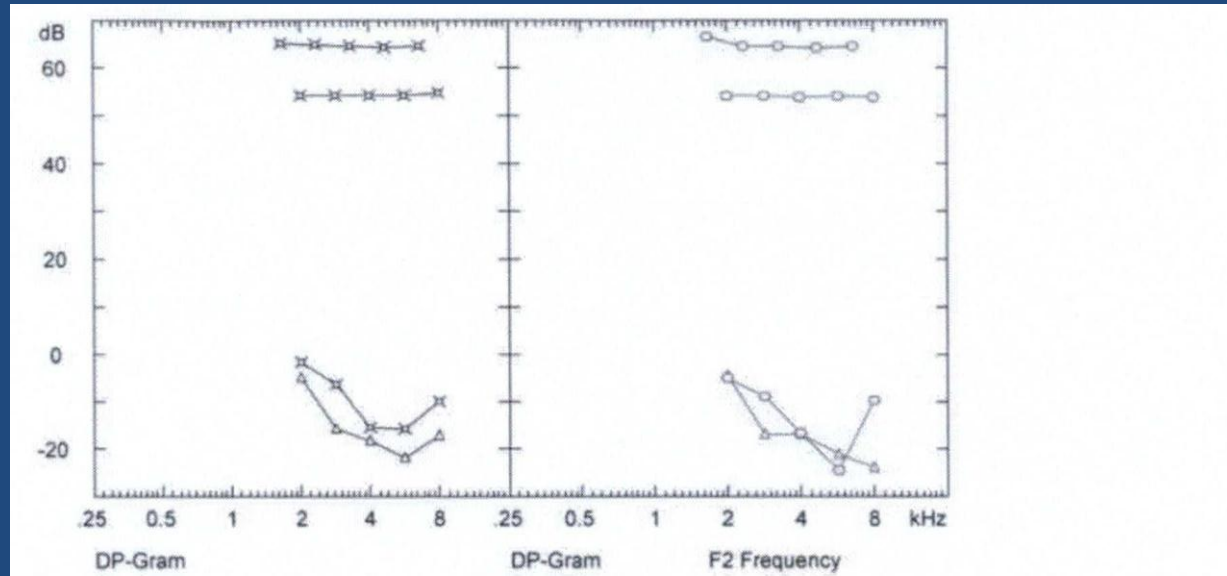


Left ear results similar, no sign of ANSD (no cochlear microphonic)

Absent DPOAEs in both ears



http://www.oae.it/old/guest_editorials/2004/01_2004.html



jfs4548 -:

Right: 27-Sep-12: -: 1.5-8kHz Diag Test High Noise: 12I27D02.OAE

L1(dB)	L2(dB)	F1(Hz)	F2(Hz)	GM(Hz)	DP(dB)	NF(dB)	DP-NF(dB)
64.7	54.1	6563	8016	7253	-9.7	-24.1	14.4
64.2	54.2	4641	5672	5130	-24.7	-21.2	-3.5
64.6	54.1	3281	4008	3626	-16.8	-17.1	0.3
64.6	54.3	2320	2836	2565	-8.9	-17.1	8.2
66.5	54.3	1641	1992	1808	-5.1	-4.6	-0.5

jfs4548 -:

Left: 27-Sep-12: -: 1.5-8kHz Diag Test High Noise: 12I27D03.OAE

L1(dB)	L2(dB)	F1(Hz)	F2(Hz)	GM(Hz)	DP(dB)	NF(dB)	DP-NF(dB)
64.6	54.9	6563	8016	7253	-10.0	-17.4	7.4
64.2	54.2	4641	5672	5130	-15.9	-22.1	6.2
64.7	54.2	3281	4008	3626	-15.6	-18.5	2.9
64.8	54.3	2320	2836	2565	-6.5	-15.8	9.3
65.0	54.2	1641	1992	1808	-1.8	-5.1	3.3

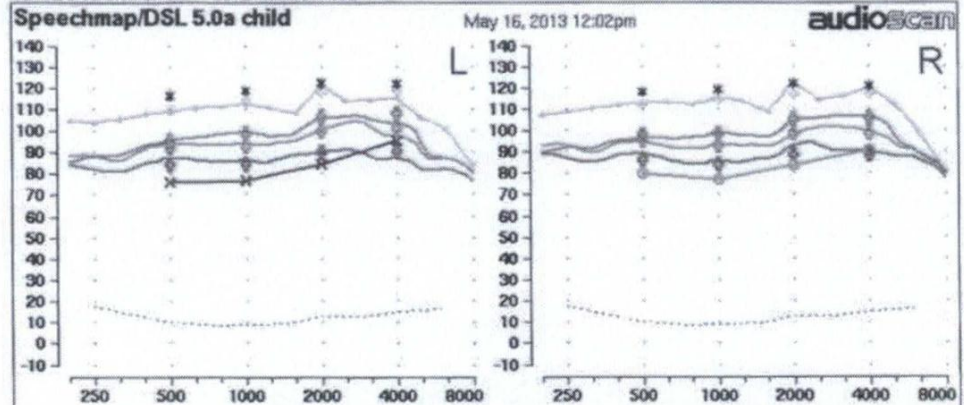
Summary of results from diagnostic audiology appointments (NR=no response)

Toneburst ABR	500 Hz	1000 Hz	2000 Hz	4000 Hz
Left AC	65	65	70	70
Right AC	65	65	70	85
BC Left & Right	NR 60		NR 60	
Click ABR				
Left & Right AC	Small wave V present; no evidence of ANSD			
Tympanometry	Type A tympanograms Absent ipsilateral acoustic reflexes			
DPOAEs	Absent left and right whenever tested			

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





Audiometry					Audiometry						
Age	31 months	Test	Stimulus	Level	SII	Age	31 months	Test	Stimulus	Level	SII
Xdcr	Insert+Foam	1	Speech-std(1)	Soft (55)	41	Xdcr	Insert+Foam	1	Speech-std(1)	Soft (55)	45
UCL	Average	2	Speech-std(1)	Avg (65)	60	UCL	Average	2	Speech-std(1)	Avg (65)	61
RECD	Measured	3	Speech-std(1)	Loud (75)	65	RECD	Measured	3	Speech-std(1)	Loud (75)	69
BCT	N/A	4	MPO	90	N/A	BCT	N/A	4	MPO	90	N/A
Unaided avg (65)					0	Unaided avg (65)					0

Speechmap/DSL 5.0a child										
Left						Right				
	250	500	750	1k	1k5	2k	3k	4k	6k	
Entered UCL										
Test 1	82	88	86	86	88	90	90	88	83	88
Test 2	88	94	94	94	95	101	104	98	87	93
Test 3	88	96	98	98	98	106	107	104	88	92
Test 4	104	109	111	112	109	122	114	115	102	109
Entered HL		65		65		70		85		65
HA-2 RECD	0	5	9	11	10	9	11	9	10	6
Entered BCT										
nHL to eHL	30	20	17	15	12	10	7	5	5	30

Audiometry					Audiometry						
Age	31 months	Test	Stimulus	Level	SII	Age	31 months	Test	Stimulus	Level	SII
Xdcr	Insert+Foam	1	Speech-std(1)	Soft (55)	41	Xdcr	Insert+Foam	1	Speech-std(1)	Soft (55)	45
UCL	Average	2	Speech-std(1)	Avg (65)	60	UCL	Average	2	Speech-std(1)	Avg (65)	61
RECD	Measured	3	Speech-std(1)	Loud (75)	65	RECD	Measured	3	Speech-std(1)	Loud (75)	69
BCT	N/A	4	MPO	90	N/A	BCT	N/A	4	MPO	90	N/A
Unaided avg (65)					0	Unaided avg (65)					0

Example of real ear measures

Measured RECDs

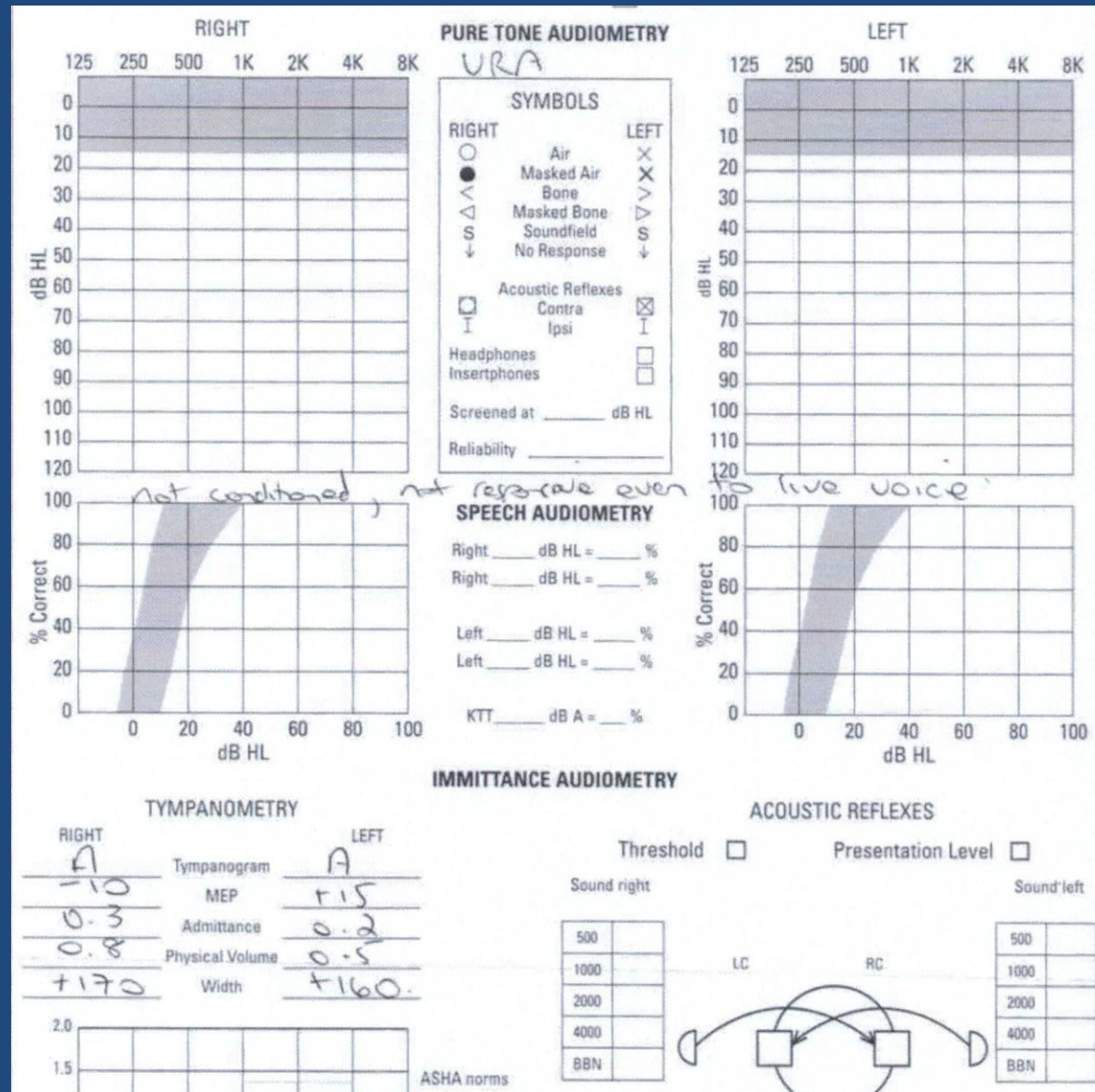
Standard speech input at soft, average and loud and MPO check with 90 dB signal

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

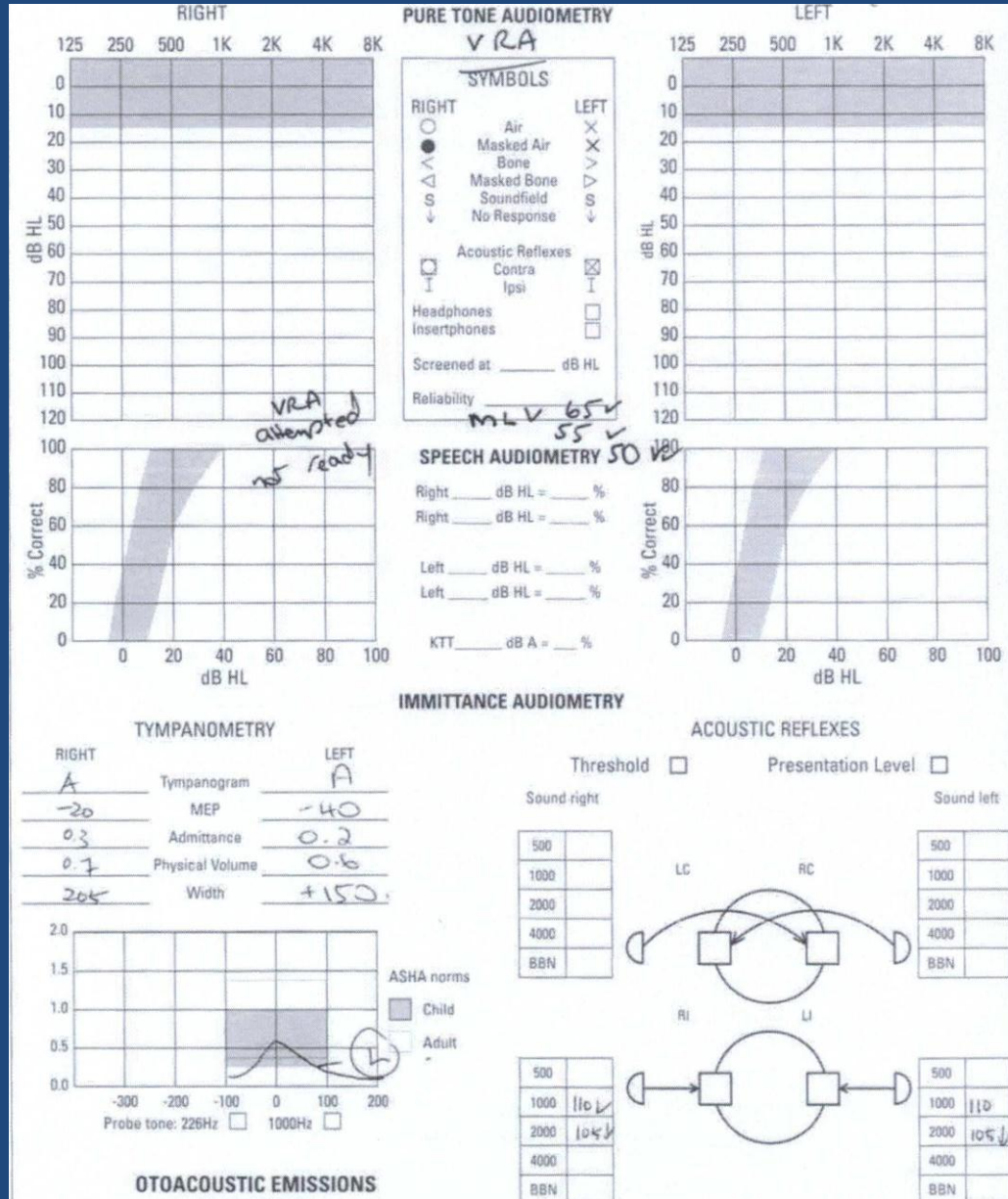
VRA at 8 months



“not conditioned,
not responsive even
to live voice”



VRA at 9 months

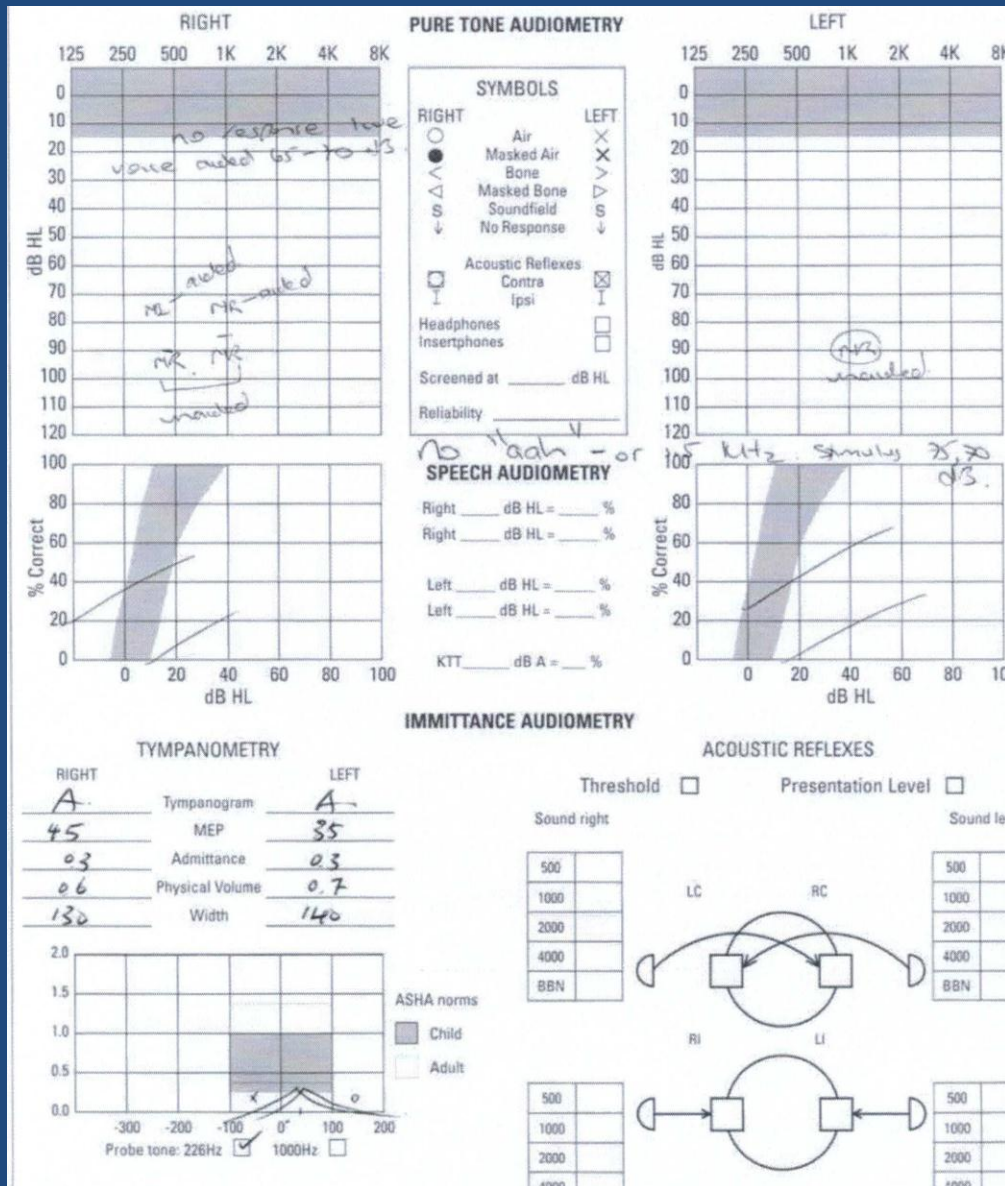


Still not ready for any frequency specific testing

Monitored live voice testing at 50dBHL?

Not very consistent with ABR thresholds

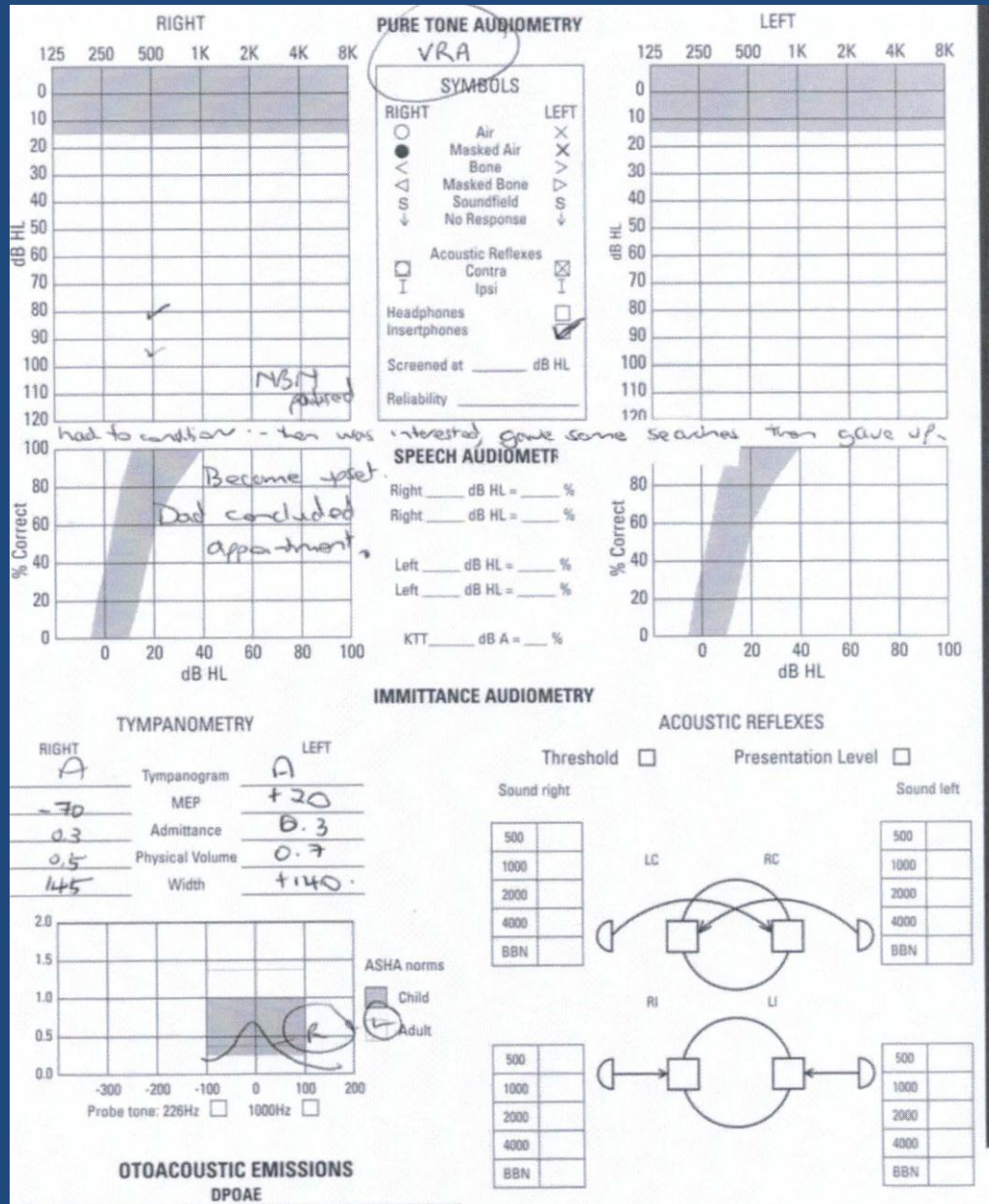
VRA at 10 months



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

Everyone getting a bit stressed

VRA at 11 months

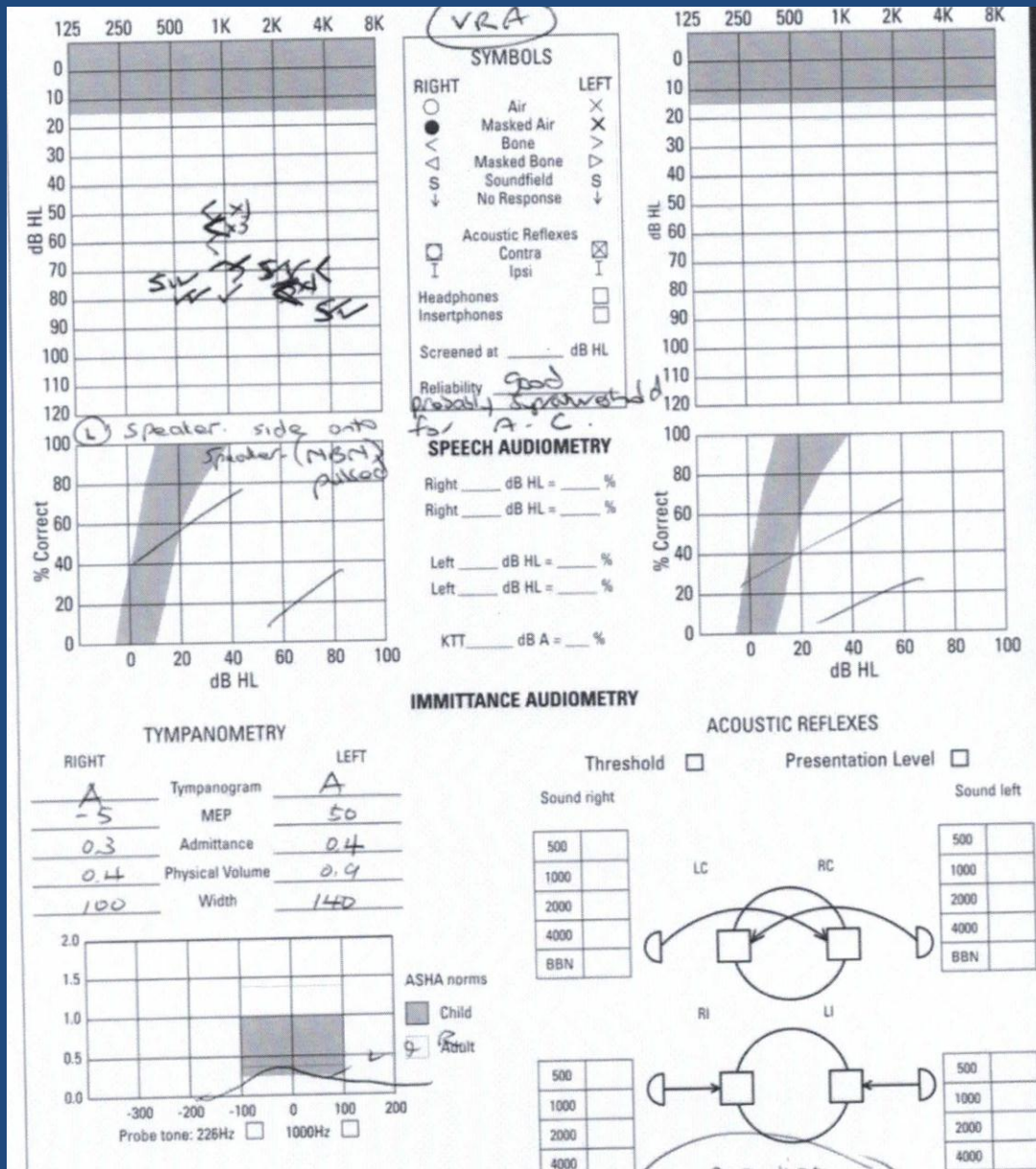


“Had to condition, then was interested, gave some searches, then gave up”

“child became upset, dad concluded appointment”

Discussions begun about repeating ABR

VRA at 15 months



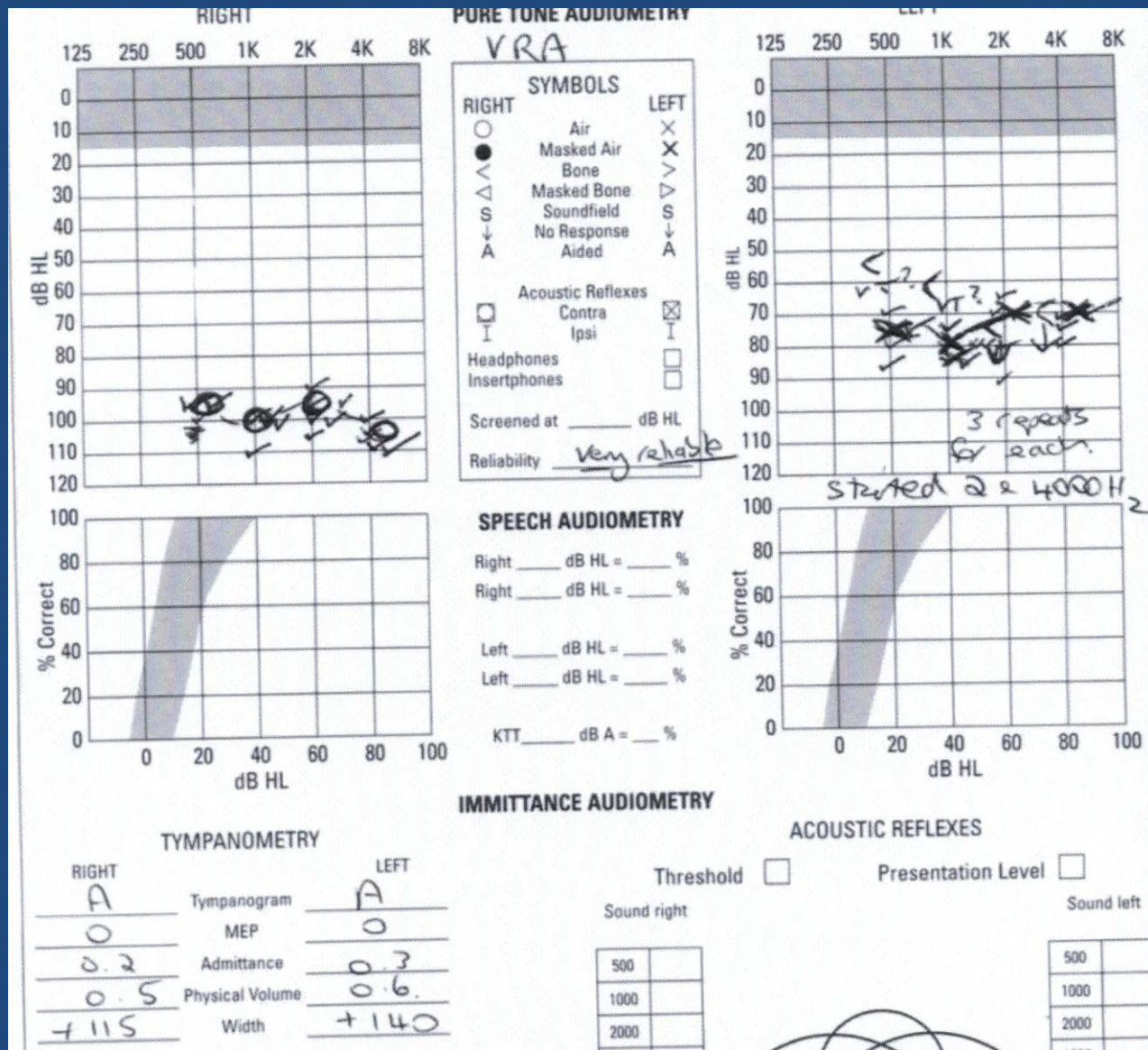
4 appointments later

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

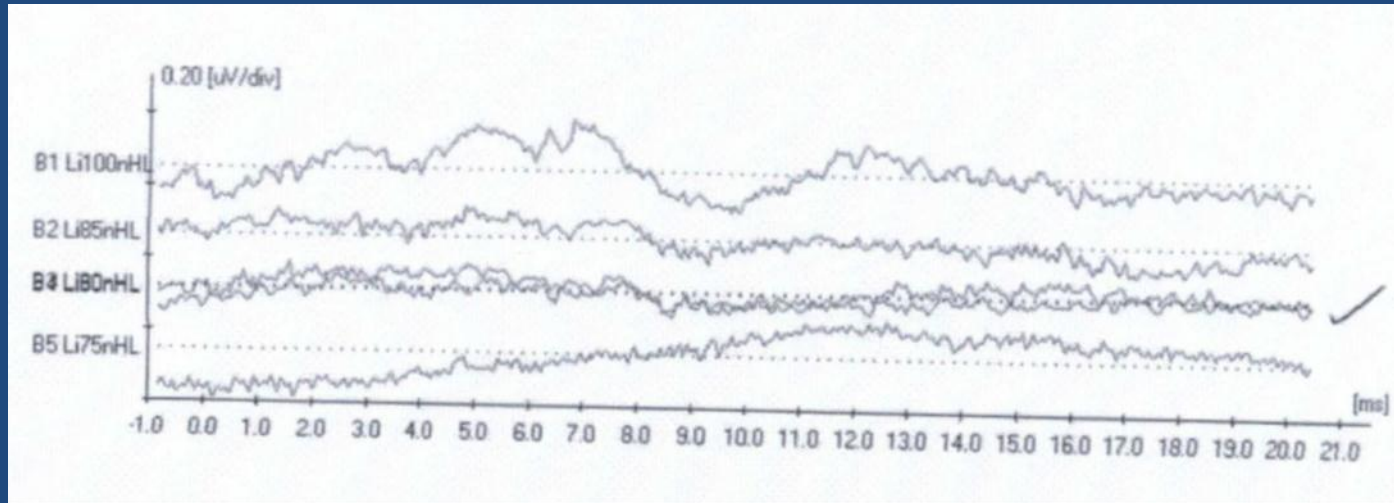
Everyone including parents want to have repeat ABR to check on thresholds

Cortical Evoked Potential Testing

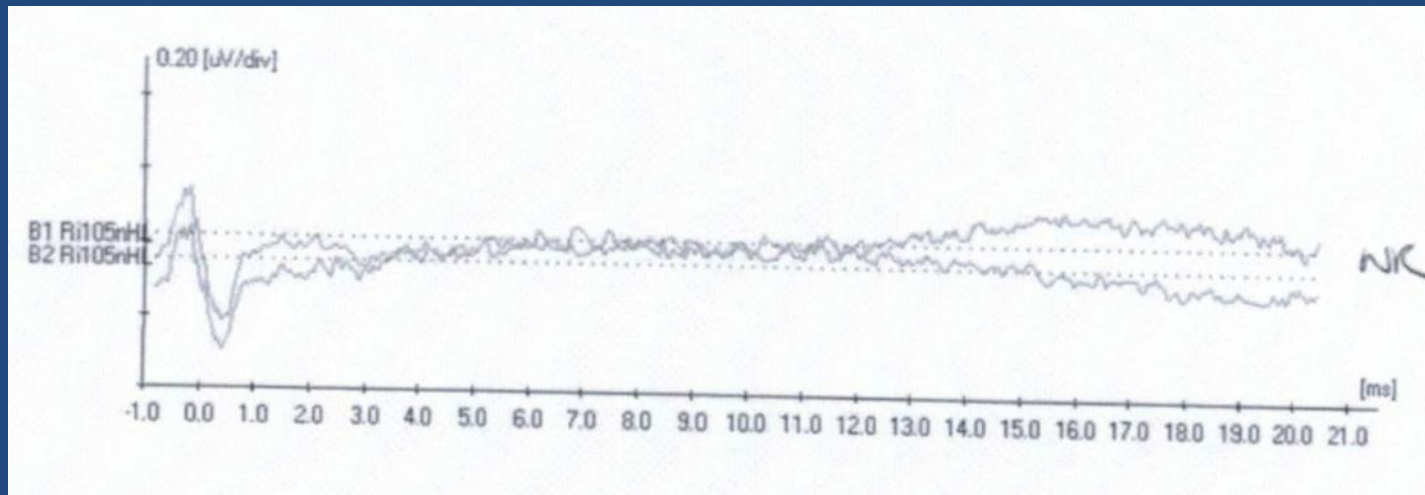
- Child was non-compliant and session abandoned, parents very keen to repeat ABR



GA (General anaesthetic) ABR

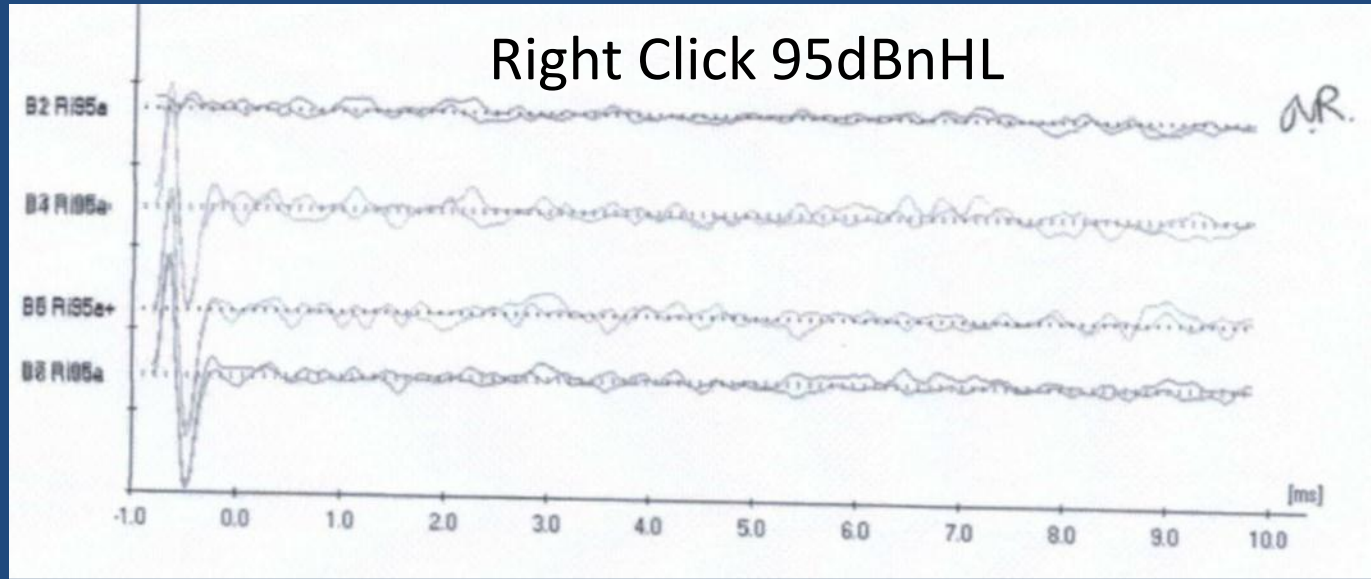


Left ear
4kHz 80 dBnHL
(response at 70
dBnHL at initial
test)

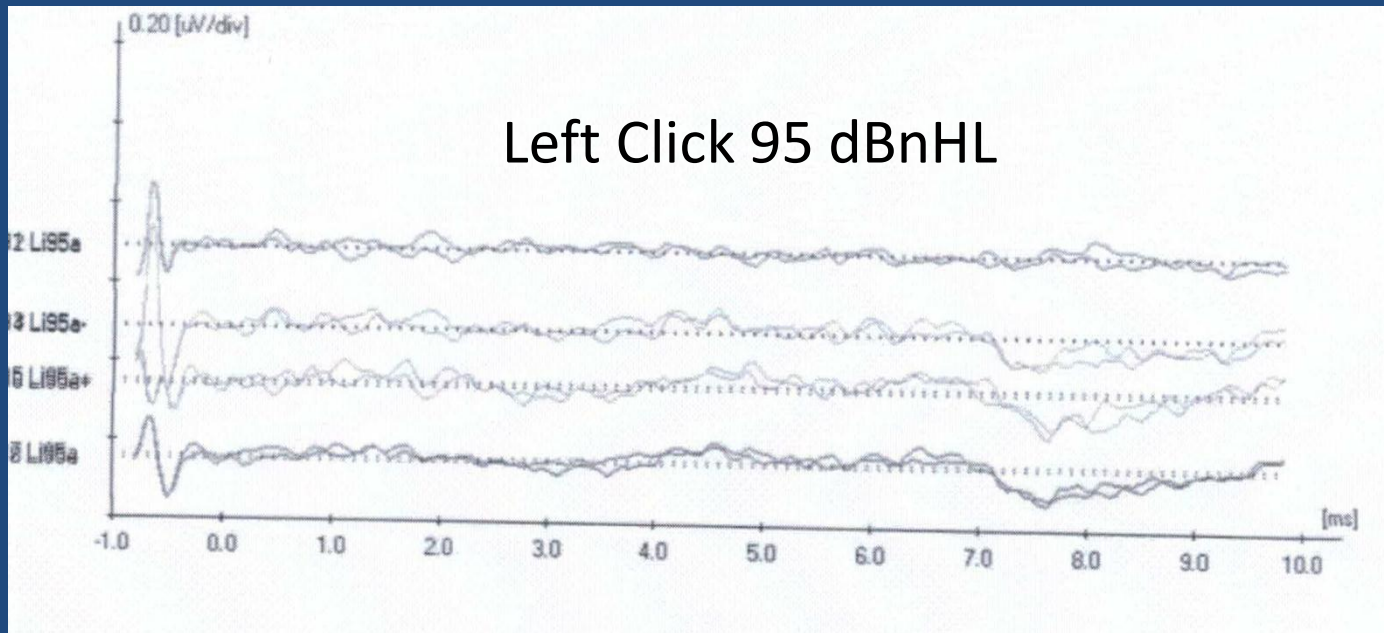


Right ear
NR at 105 dBnHL
(response at 85
dBnHL previously)

GA ABR

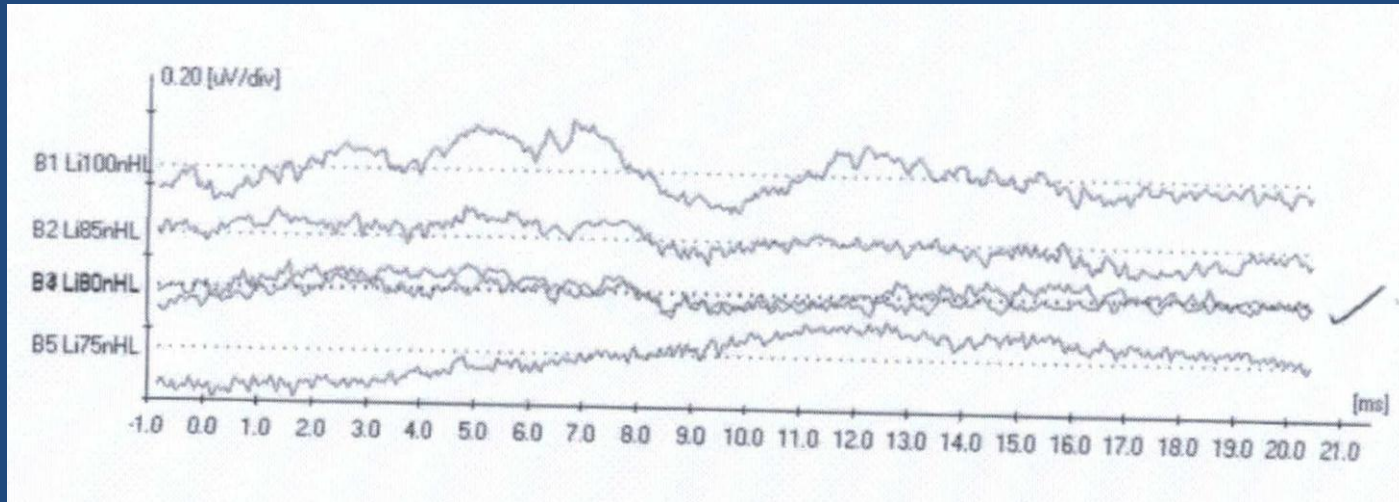


Subtracted
Rarefaction
Condensation
Alternating
Right NR

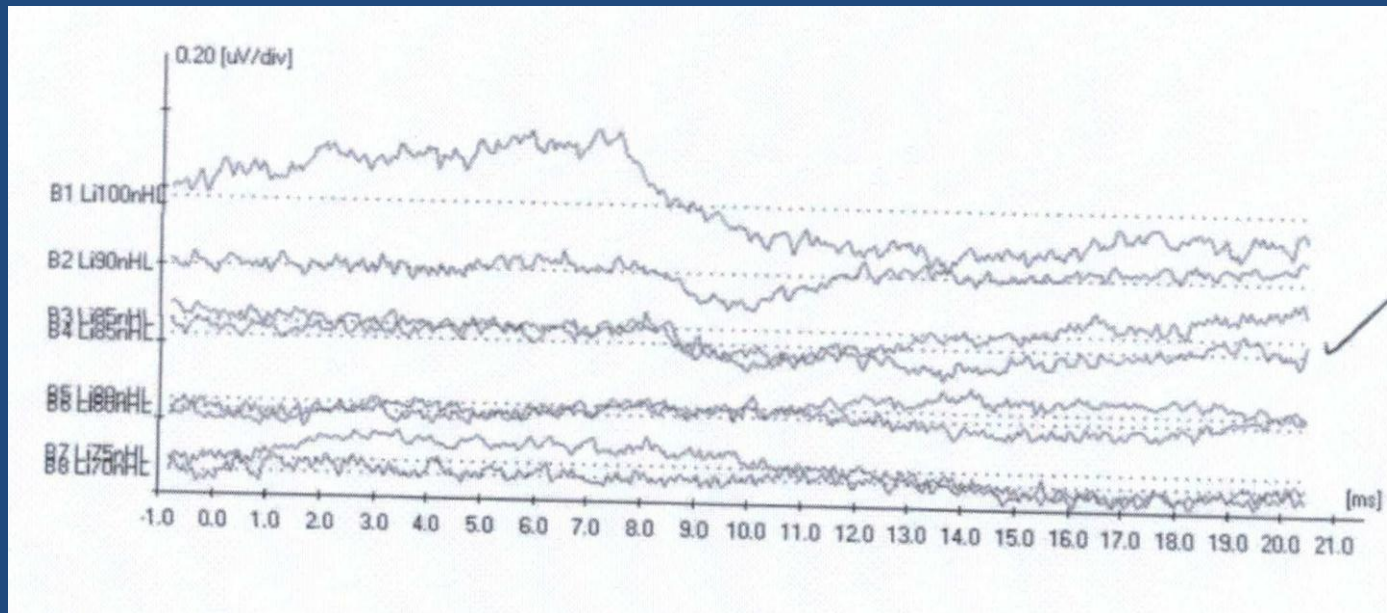


Left Small
wave V at 95
dBnHL
No CM present

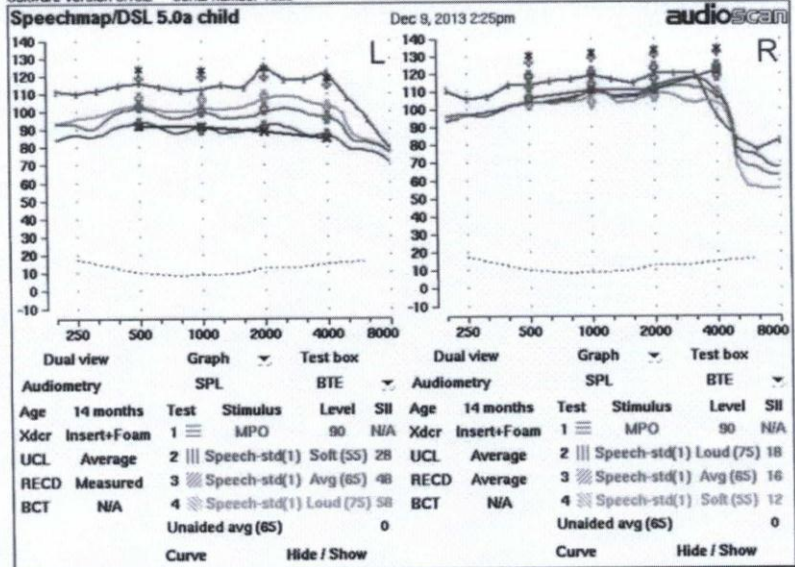
GA ABR



Left 4kHz
80 dBnHL
(previous test
threshold=70)



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



Speechmap/DSL 5.0a child

Left											Right										
250	500	750	1k	1k5	2k	3k	4k	6k	250	500	750	1k	1k5	2k	3k	4k	6k				
Entered UCL											Entered UCL										
Test 1	110	116	112	112	114	126	118	122	89	Test 1	106	114	117	119	116	120	121	96	78		
Test 2	87	94	88	91	89	82	89	87	78	Test 2	97	104	108	110	108	113	118	110	75		
Test 3	93	102	96	89	96	100	100	97	83	Test 3	97	104	105	111	107	111	113	108	68		
Test 4	96	103	100	102	102	107	108	104	85	Test 4	97	104	104	108	105	108	105	104	56		
Entered HL	75	80	70	70	70	70	70	70	70	Entered HL	85	100	95	105	105	105	105	105	105		
HA-2 RECD	6	12	9	11	13	13	14	13	5	HA-2 RECD	3	6	8	10	10	10	11	15	16		
Entered BCT										Entered BCT											
nHL to eHL	30	20	17	15	12	10	7	5	5	nHL to eHL	30	20	17	15	12	10	7	5	5		

Add Description

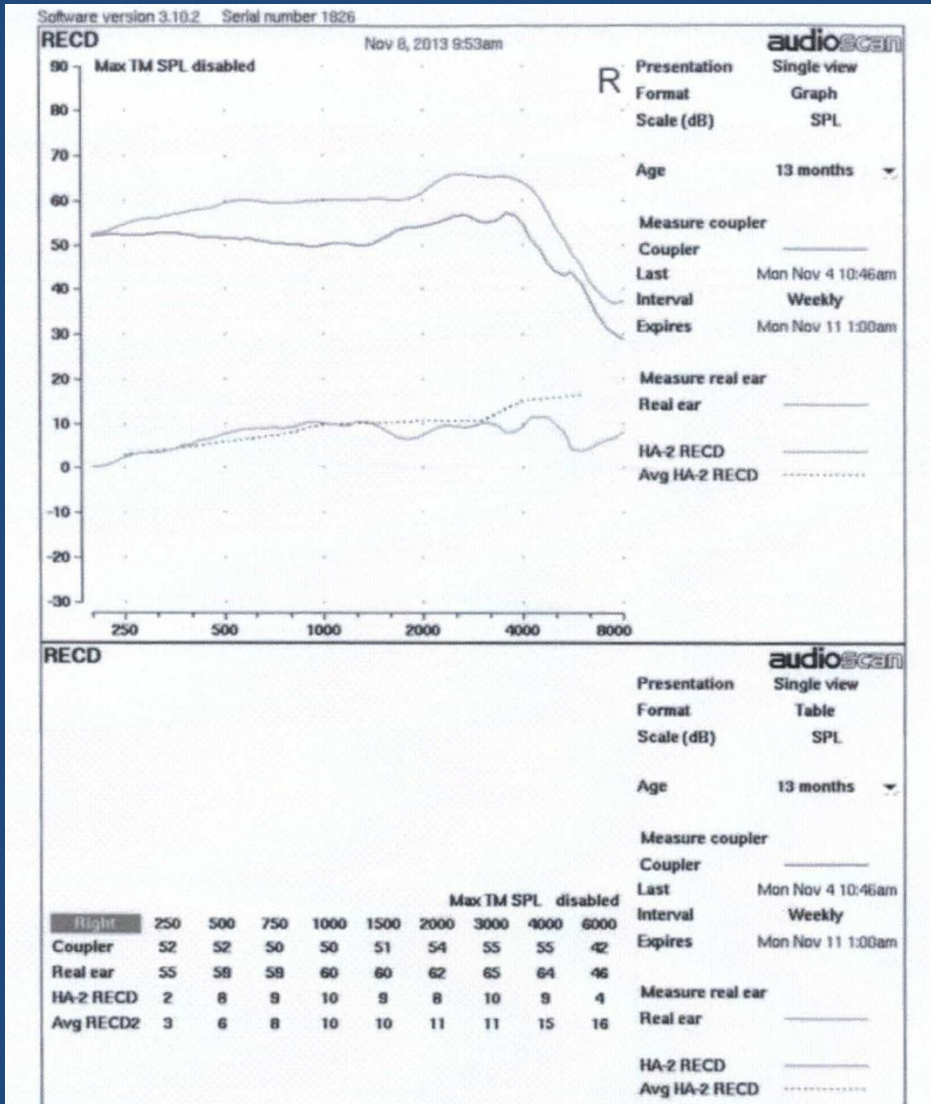
(R) - Naida IX UP

Real Ear Measures

Re-fitted for right profound loss

Naida IX UP





Measured RECDs



Upset for (L) RECD (Type A
 tympanograms both sides)

with inserts.
 used (R) & (L)

Confirmed Genetic Cause of Hearing Loss



Detected mutations

Sample no:	Gene	Exon	Nucleotide change	Amino acid change	Genotyping results	Zygoty	Type of variation	Verified by sequencing
HF0890	GJB2 (Connexin2 6)	ex2	c.283G>A	p.Val95Met	AG/CT	HET	Mut	+
HF0890	GJB2 (Connexin2 6)	ex2	c.35delG	p.Gly12fs	GT/AC	HET	Mut	+

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



