

# CHALLENGING HEARING AID FITTING PROGRESSIVE HEARING LOSS

## DISCLOSURES

- Manager of Audiology @ Hackensack University Medical Center
- COO @ Hackensack Audiology and Hearing Aid Associates

## HEALTH HISTORY

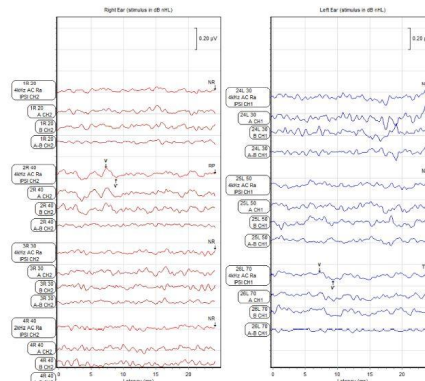
- Pregnancy was unremarkable.
- Birth was unremarkable on 12/29/2024.
- Did not pass newborn hearing screening @ outside facility.

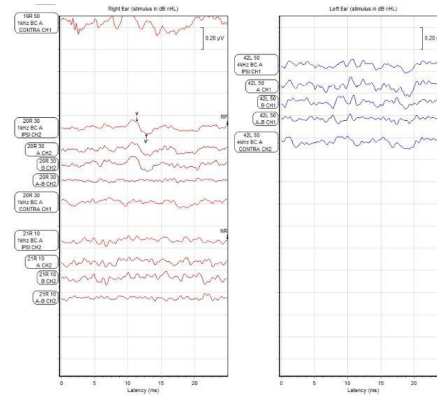
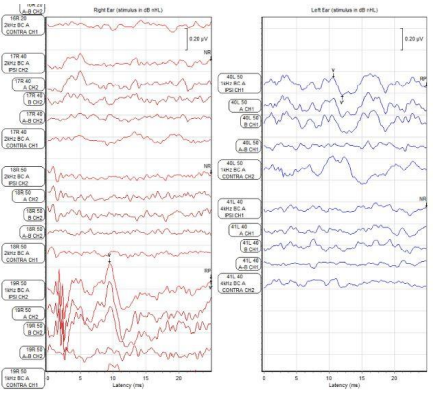
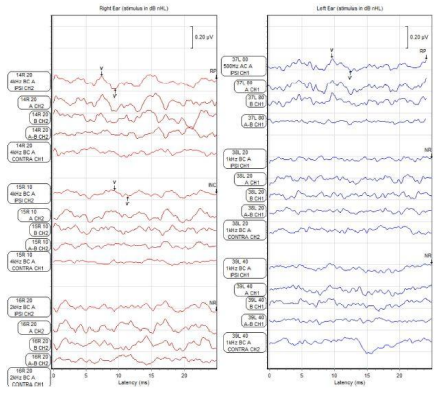
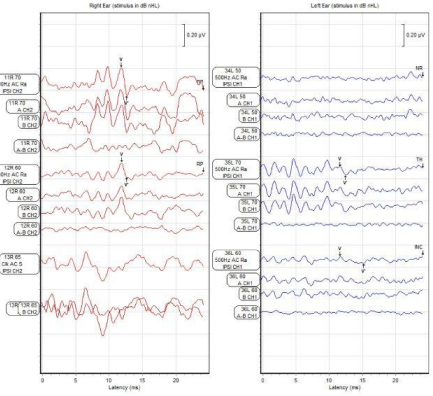
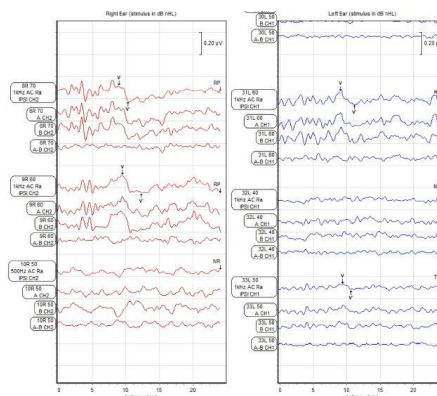
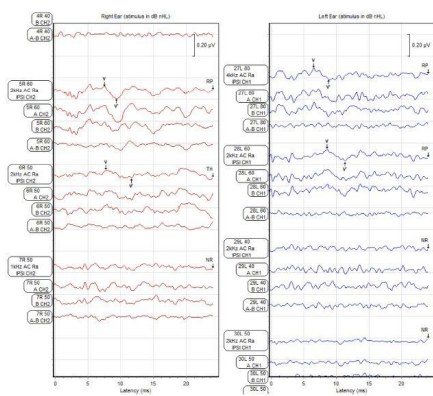
## HEALTH HISTORY CONTINUED

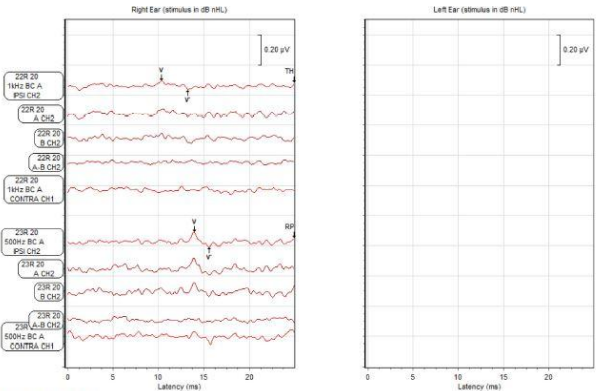
- Re-admitted on day 5 of life for hyperbilirubinemia requiring phototherapy.
- Suspected cCMV - IgG and Toxoplasma studies were negative.
- No family history of childhood onset hearing loss.

## SEEKING SECOND OPINION

- 3 month old had diagnostic ABR at Birthing Facility on 3/7/2025.
- Right ear ABR demonstrated mild SNHL.
- Left ear ABR demonstrated a moderate SNHL.
- Family did not seek amplification yet



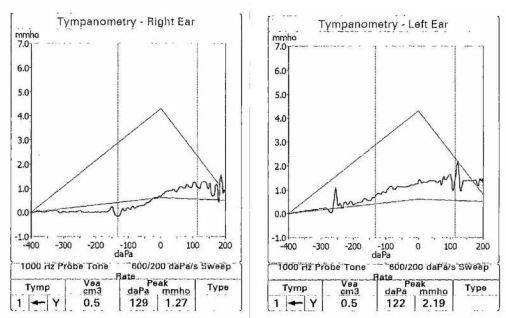




ABR WAVEFORM DATA		Corr. Coef.	RN (µV)	SNR	I	F	II	III	IV	V	VI	Lat (ms)	Amplitude (µV)	Ratio	SE
1 R 20 CH2	0.007	---	---	---	---	---	---	---	---	---	---	---	---	---	---
2 R 40 CH2	0.009	3.47	---	---	---	---	---	---	---	---	---	7.33	8.89	---	0.09
3 R 60 CH2	0.009	---	---	---	---	---	---	---	---	---	---	---	---	---	---
4 R 80 CH2	0.010	---	---	---	---	---	---	---	---	---	---	---	---	---	---
5 R 100 CH2	0.018	2.10	---	---	---	---	---	---	---	---	---	7.53	9.45	---	0.11
6 R 120 CH2	0.011	2.96	---	---	---	---	---	---	---	---	---	7.74	11.70	---	0.08
7 R 140 CH2	0.013	---	---	---	---	---	---	---	---	---	---	---	---	---	---
8 R 160 CH2	0.011	4.00	---	---	---	---	---	---	---	---	---	8.99	10.35	---	0.14
9 R 180 CH2	0.016	3.24	---	---	---	---	---	---	---	---	---	9.51	12.32	---	0.16
10 R 200 CH2	0.017	---	---	---	---	---	---	---	---	---	---	---	---	---	---
11 R 220 CH2	0.025	3.46	---	---	---	---	---	---	---	---	---	11.80	12.64	---	0.25
12 R 240 CH2	0.012	3.37	---	---	---	---	---	---	---	---	---	11.91	12.64	---	0.13
13 R 260 CH2	0.046	---	---	---	---	---	---	---	---	---	---	---	---	---	---
14 R 280 CH2	0.014	---	---	---	---	---	---	---	---	---	---	---	---	---	---
14 R 300 CH2	0.016	2.81	---	---	---	---	---	---	---	---	---	7.40	9.48	---	0.11
15 R 320 CH2	0.007	---	---	---	---	---	---	---	---	---	---	---	---	---	---
15 R 340 CH2	0.014	1.35	---	---	---	---	---	---	---	---	---	9.38	11.35	---	0.05
16 R 360 CH2	0.012	---	---	---	---	---	---	---	---	---	---	---	---	---	---
17 R 380 CH2	0.016	---	---	---	---	---	---	---	---	---	---	---	---	---	---
18 R 400 CH2	0.021	---	---	---	---	---	---	---	---	---	---	---	---	---	---
18 R 420 CH2	0.012	---	---	---	---	---	---	---	---	---	---	---	---	---	---
19 R 440 CH2	0.013	---	---	---	---	---	---	---	---	---	---	---	---	---	---
19 R 460 CH2	0.031	1.51	---	---	---	---	---	---	---	---	---	9.48	24.90	---	0.15
20 R 480 CH2	0.011	---	---	---	---	---	---	---	---	---	---	---	---	---	---
20 R 500 CH2	0.009	4.16	---	---	---	---	---	---	---	---	---	11.35	12.81	---	0.10
21 R 520 CH2	0.008	---	---	---	---	---	---	---	---	---	---	---	---	---	---
22 R 540 CH2	0.007	2.61	---	---	---	---	---	---	---	---	---	10.31	13.23	---	0.05
23 R 560 CH2	0.010	---	---	---	---	---	---	---	---	---	---	---	---	---	---
23 R 580 CH2	0.011	3.93	---	---	---	---	---	---	---	---	---	13.96	15.62	---	0.09
24 L 60 CH2	0.012	---	---	---	---	---	---	---	---	---	---	---	---	---	---
24 L 80 CH2	0.015	---	---	---	---	---	---	---	---	---	---	---	---	---	---
24 L 70 CH2	0.009	3.04	---	---	---	---	---	---	---	---	---	7.22	9.30	---	0.07

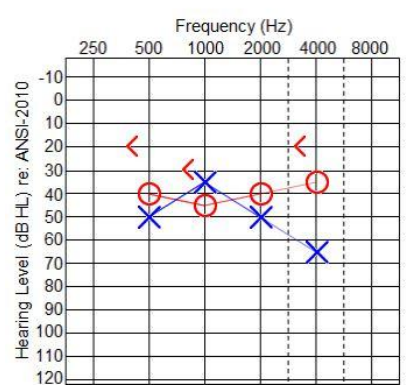
ABR WAVEFORM DATA		Corr. Coef.	RN (µV)	SNR	I	F	II	III	IV	V	VI	Lat (ms)	Amplitude (µV)	Ratio	SE
27 L 60 CH2	0.012	3.31	---	---	---	---	---	---	---	---	---	8.49	8.78	---	0.10
28 L 80 CH2	0.011	4.14	---	---	---	---	---	---	---	---	---	8.58	11.28	---	0.11
29 L 100 CH2	0.014	---	---	---	---	---	---	---	---	---	---	---	---	---	---
30 L 120 CH2	0.007	---	---	---	---	---	---	---	---	---	---	---	---	---	---
31 L 140 CH2	0.015	2.79	---	---	---	---	---	---	---	---	---	9.10	11.18	---	0.12
32 L 160 CH2	0.009	---	---	---	---	---	---	---	---	---	---	---	---	---	---
33 L 180 CH2	0.007	2.47	---	---	---	---	---	---	---	---	---	9.41	10.66	---	0.05
34 L 200 CH2	0.011	---	---	---	---	---	---	---	---	---	---	---	---	---	---
35 L 220 CH2	0.007	3.33	---	---	---	---	---	---	---	---	---	11.60	12.43	---	0.07
36 L 240 CH2	0.008	2.61	---	---	---	---	---	---	---	---	---	11.60	15.14	---	0.06
37 L 260 CH2	0.010	3.71	---	---	---	---	---	---	---	---	---	9.62	12.43	---	0.11
38 L 280 CH2	0.012	---	---	---	---	---	---	---	---	---	---	---	---	---	---
39 L 300 CH2	0.016	---	---	---	---	---	---	---	---	---	---	---	---	---	---
40 L 320 CH2	0.015	4.35	---	---	---	---	---	---	---	---	---	10.62	11.98	---	0.15
41 L 340 CH2	0.013	---	---	---	---	---	---	---	---	---	---	---	---	---	---
42 L 360 CH2	0.020	---	---	---	---	---	---	---	---	---	---	---	---	---	---
43 L 380 CH2	0.018	---	---	---	---	---	---	---	---	---	---	---	---	---	---
44 L 400 CH2	0.020	---	---	---	---	---	---	---	---	---	---	---	---	---	---

Test date: 4/23/2025  
TymStar Pro, GS0082585



- No point of maximal tympanometric peak pressure or static compliance.
- OAEs not performed b/c of hearing loss, abnormal MEA, and time constraints.

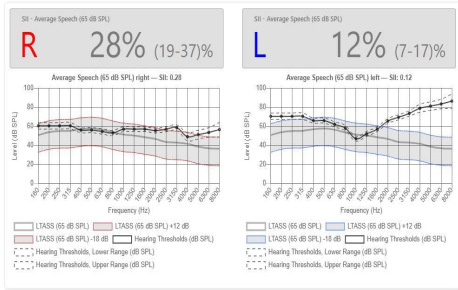
### ESTIMATED AUDIOGRAM



### RECOMMENDATIONS - 4/23/2025

- F/U with ENT.
- Trial with HAs if expected to be long-standing or not ready to be managed medically/surgically.
  - Unaided SII - 28% right and 12% left (65 dB SPL)
- ABR re-evaluation in 3 months or per ENT.

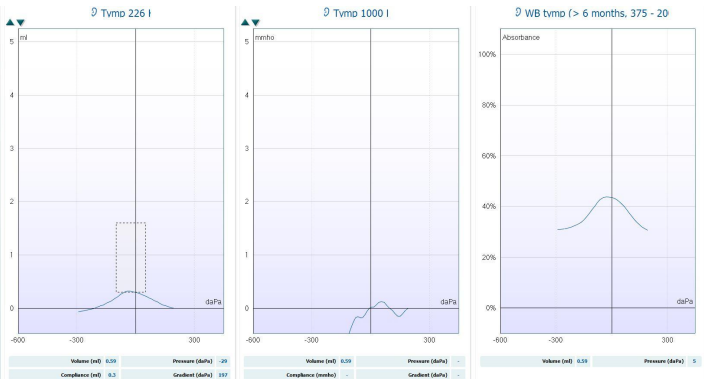
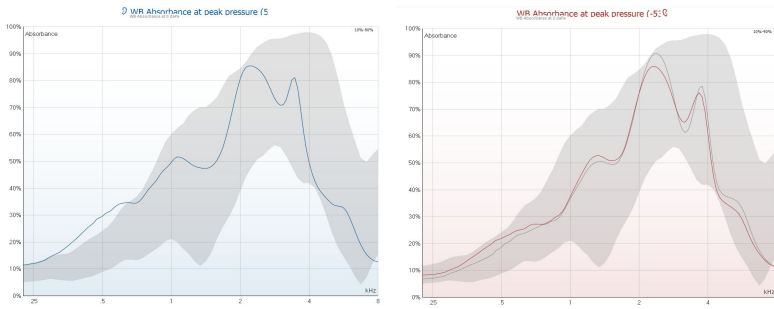
# UNAIDED SII



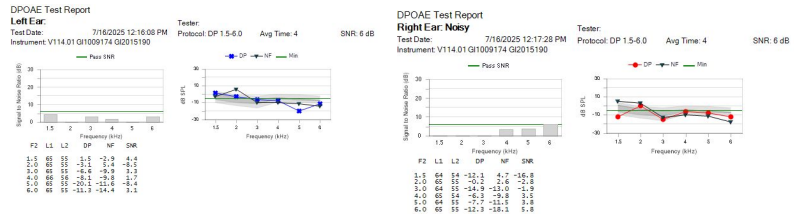
# Re-evaluation 7/2025

- Re-assessment prior to getting ENT clearance for HAs

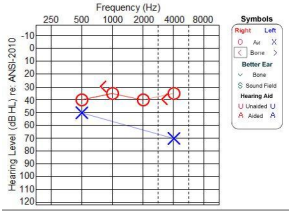
# WIDE BAND ABSORBANCE AT PEAK PRESSURE



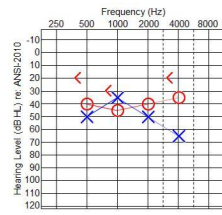
# OTOACOUSTIC EMISSIONS



# Estimated Audiogram 7/2025

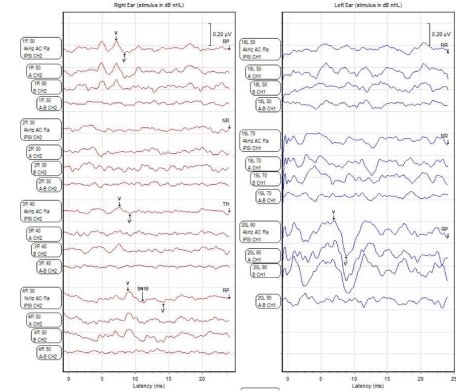


# Estimated Audiogram 3/2025

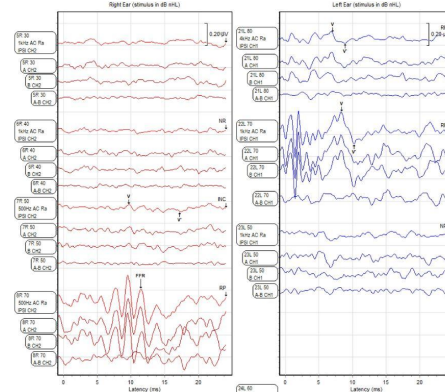


# RE-EVALUATION 7/16/2025

- Recommendations
  - Trial with amplification.
  - Genetics consultation

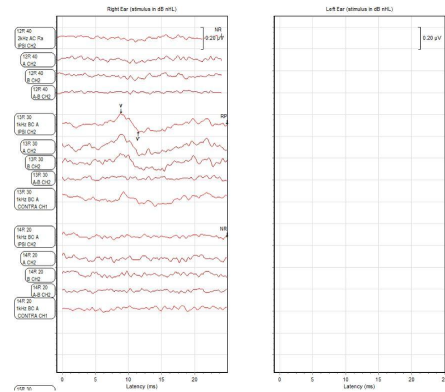
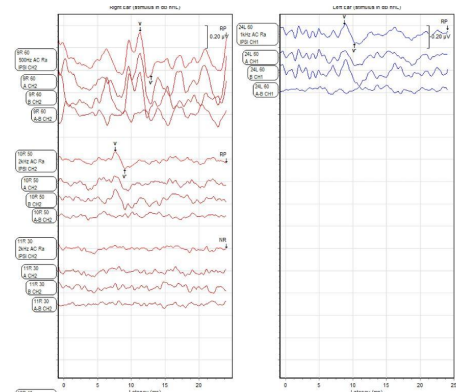


		I	II	III	IV	V	Time (ms)
1	R 50 CH2	---	---	---	---	---	7.12
3	R 40 CH2	---	---	---	---	---	7.44
4	R 30 CH2	---	---	---	---	---	8.89
7	R 50 CH2	---	---	---	---	---	9.72
9	R 40 CH2	---	---	---	---	---	11.38
10	R 30 CH2	---	---	---	---	---	7.64
13	R 50 CH1	---	---	---	---	---	8.85
16	R 40 CH1	---	---	---	---	---	7.40
18	R 30 CH1	---	---	---	---	---	6.91
20	L 50 CH1	---	---	---	---	---	7.01
22	L 40 CH1	---	---	---	---	---	8.37
24	L 30 CH1	---	---	---	---	---	8.78



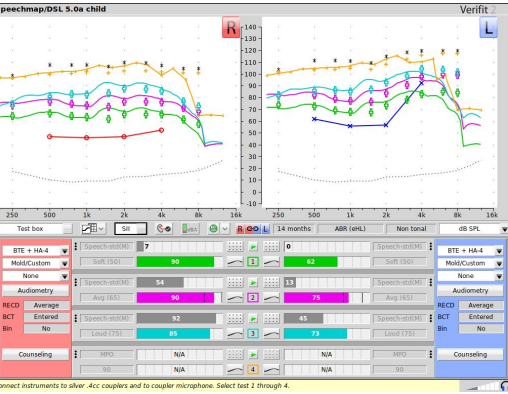
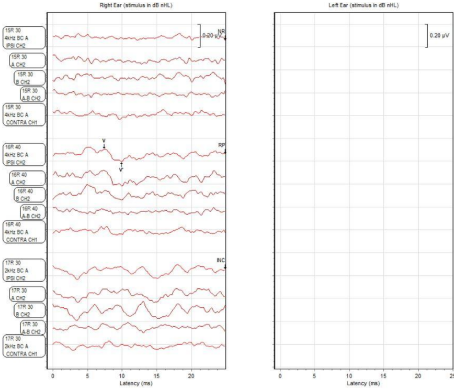
AEP TEST RESULTS - Version of Software When Collected: 6.10.4

	Stimulus	Gain	Latency	Amplitude	SNR	Pass	Fail	Pass	Fail
1	R 50 CH2	---	---	---	---	---	---	---	---
3	R 40 CH2	---	---	---	---	---	---	---	---
4	R 30 CH2	---	---	---	---	---	---	---	---
7	R 50 CH2	---	---	---	---	---	---	---	---
9	R 40 CH2	---	---	---	---	---	---	---	---
10	R 30 CH2	---	---	---	---	---	---	---	---
13	R 50 CH1	---	---	---	---	---	---	---	---
16	R 40 CH1	---	---	---	---	---	---	---	---
18	R 30 CH1	---	---	---	---	---	---	---	---
20	L 50 CH1	---	---	---	---	---	---	---	---
22	L 40 CH1	---	---	---	---	---	---	---	---
24	L 30 CH1	---	---	---	---	---	---	---	---



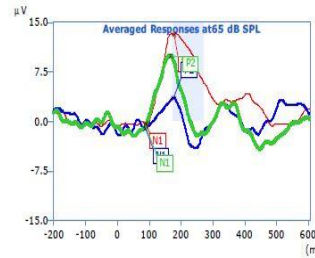
# FITTED WITH OTICON PX BTE HEARING AIDS

- Fitting to DSL 5.0 using simulated real-ear.
- Functional aided testing done via aided CAEP.
  - Good aided benefit /m/, /g/, /t/ @ 65 dB SPL @ 1.5 meters



## Simulated Real-Ear

## RIGHT AIDED CAEP RESPONSE



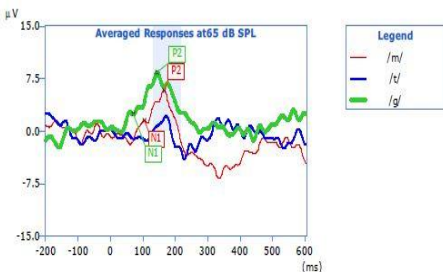
Latencies for responses at 65 dB SPL (ms)

	/m/	/t/	/g/	/s/
P1				
N1	89	86	87	
P2	176	173	167	

Were client's responses to the stimuli presented significantly different from noise? (i.e. p <= 0.05)

	/m/	/t/	/g/	/s/
5 dB SP	0.000	0.000	0.000	

## LEFT AIDED CAEP RESPONSE



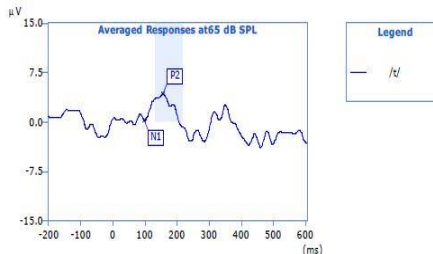
Latencies for responses at 65 dB SPL (ms)

	/m/	/t/	/g/	/s/
P1				
N1	104		68	
P2	164		143	

Were client's responses to the stimuli presented significantly different from noise? (i.e. p <= 0.05)

	/m/	/t/	/g/	/s/
5 dB SP	0.000	0.110	0.002	

## LEFT AIDED CAEP RESPONSE



Latencies for responses at 65 dB SPL (ms)

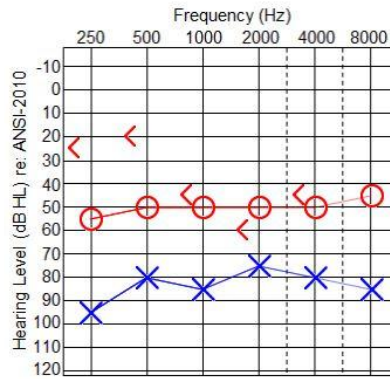
	/m/	/t/	/g/	/s/
P1				
N1		95		
P2		155		

Were client's responses to the stimuli presented significantly different from noise? (i.e. p <= 0.05)

	/m/	/t/	/g/	/s/
5 dB SP		0.001		

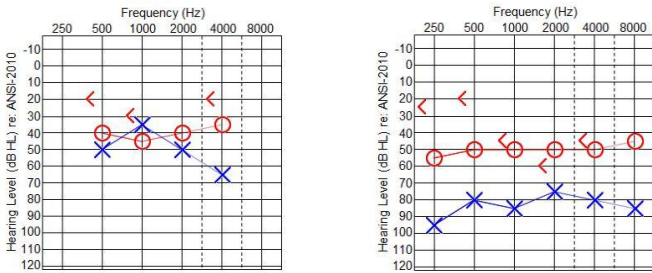
## PARENTAL CONCERN REPORTED OCT. 2025

- Parents reported that child was not as responsive to sound.
- An audiological evaluation was recommended to assess child's auditory function and needs.
- Reprogram hearing aids if change in hearing is identified.

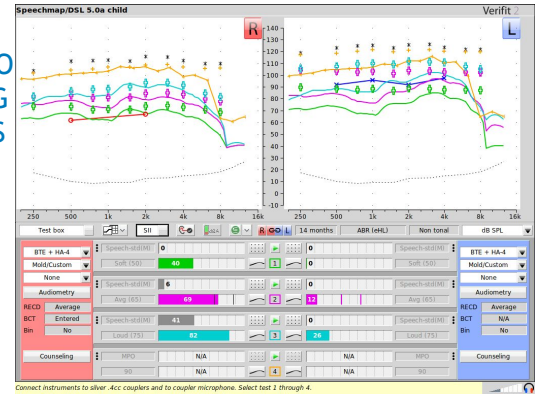


## COMPOSITE AUDIOGRAM OVER TWO SESSIONS

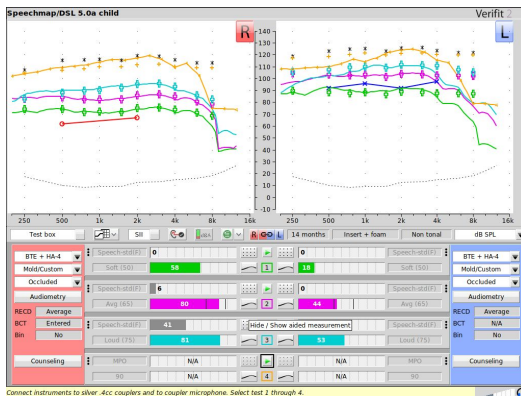
## COMPARISON FROM 3/2025 AND 12/2025



## HEARING AIDS NO LONGER HITTING DSL 5.0 TARGETS



## REPROGRAMMED HEARING AIDS



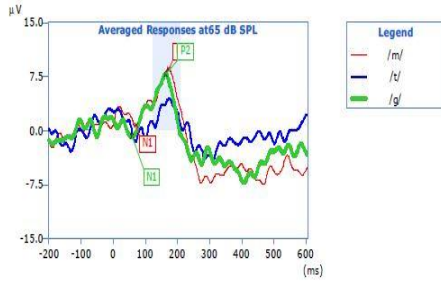
Connect instruments to silver .4cc couplers and to coupler microphone. Select test 2 through 4.

## PLAN OF ACTION

- Significant change in hearing since diagnosis.
- Cortical Auditory Evoked Potentials to confirm appropriate aided benefit
- Parents received diagnosis - Pendred syndrome

	RMSE	
	Right	Left
50 dB SPL	1.2	3.1
65 dB SPL	1.4	3.0
75 dB SPL	1.7	4.4

## RIGHT AIDED CAEP RESPONSE



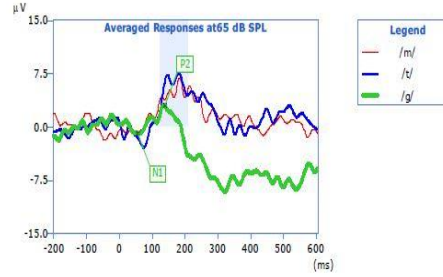
Latencies for responses at 65 dB SPL (ms)

	/m/	/t/	/g/	/s/
<b>P1</b>				
<b>N1</b>	70		57	
<b>P2</b>	172		160	

Were client's responses to the stimuli presented significantly different from noise? (i.e.  $p \leq 0.05$ )

	/m/	/t/	/g/	/s/
<b>5 dB SP</b>	<b>0.000</b>	0.068	<b>0.000</b>	

## LEFT AIDED CAEP RESPONSE



Latencies for responses at 65 dB SPL (ms)

	/m/	/t/	/g/	/s/
<b>P1</b>				
<b>N1</b>			54	
<b>P2</b>			136	

Were client's responses to the stimuli presented significantly different from noise? (i.e.  $p \leq 0.05$ )

	/m/	/t/	/g/	/s/
<b>5 dB SP</b>	0.068	0.083	<b>0.000</b>	

## OUTCOME

- Cochlear implant for the left ear because aided CAEP suggests limited benefit from conventional amplification.
- Family decided to pursue CI on the left because of the greater likelihood of decrease in hearing in the future.
- Unless there is further change to his hearing he will be bimodal

## QUESTIONS?