

CI in a young adult with mild-normal hearing thresholds

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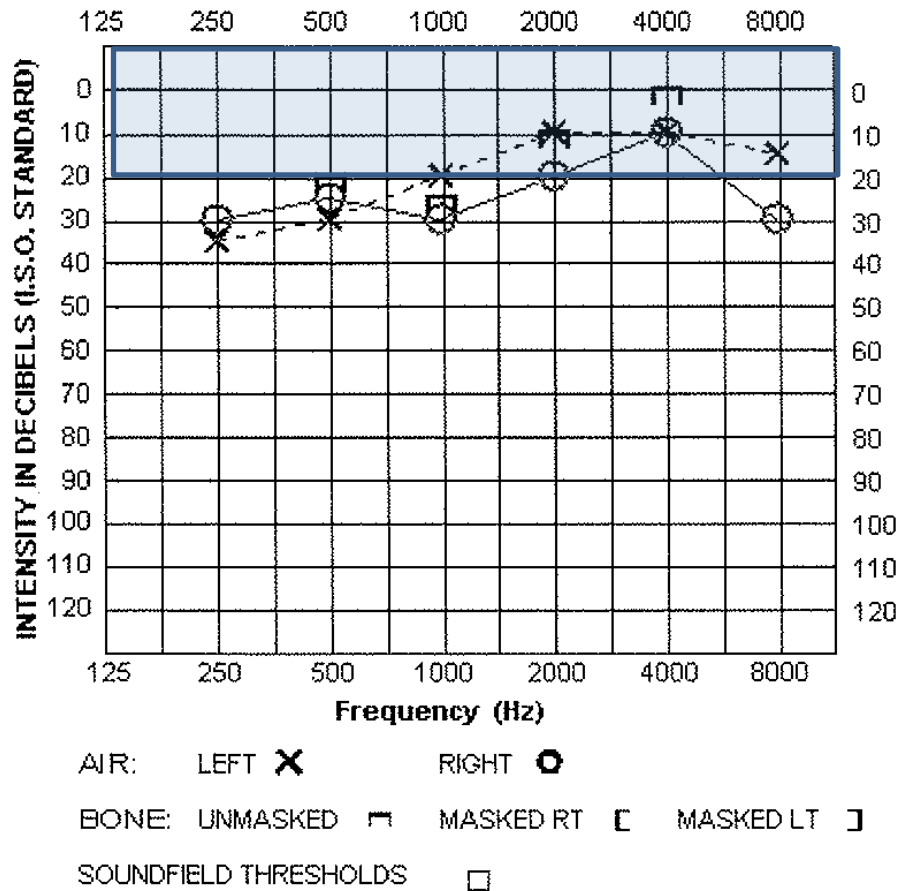
13/4/2014



S C I C

hear together

Background



Has **optic neuropathy** & vision is significantly affected as a result of a mutation in the **OPA1 gene**

Audiological assessments

- **Audiogram** only shows a mild, low & mid-frequency hearing loss
- **Tympanometry**: normal

Patient report

- Having great difficulty hearing at school & home
- Ok when talking 1:1 but prefers to write rather than converse
- Uses captions on TV
- Cannot use the phone
- Withdrawing from social situations
- Problems with hearing documented when 8 years old (now a teenager)

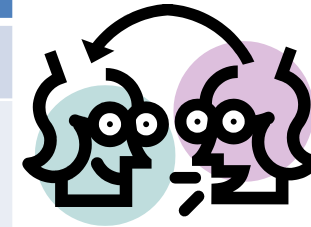
Amplification

- Tried hearing aids but of little benefit. Sounds were louder but not clearer
- Tried FM-system but of little benefit

What should we check next?

1. Speech discrimination testing:

	Left ear	Right ear
AB words	10%	20%
BKB sentences	Auditory alone = 0% Auditory-Visual = 50%	



2. Stapedial reflexes (Middle-Ear Muscle Reflexes)

	Left ear	Right ear
1 kHz tone	absent	absent



Questions:

1. What do you think the diagnosis is (why do you think the speech discrimination is worse than you would expect given the degree of the hearing loss)?
2. What test/s would you recommend next?

Suspected diagnosis:

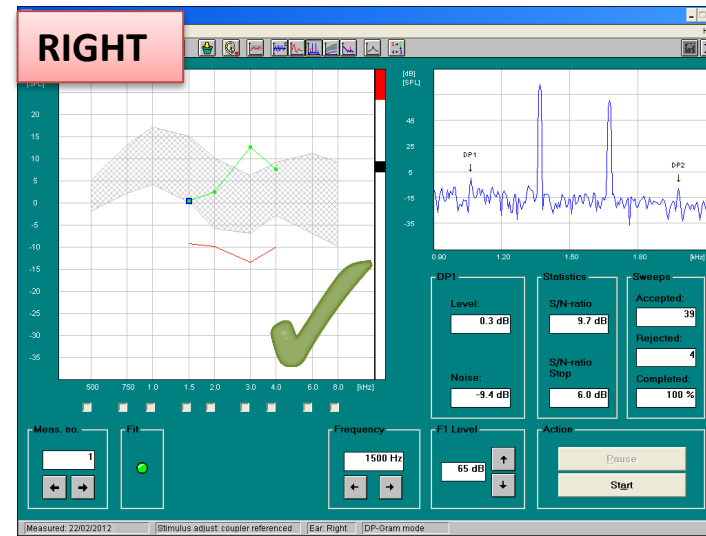
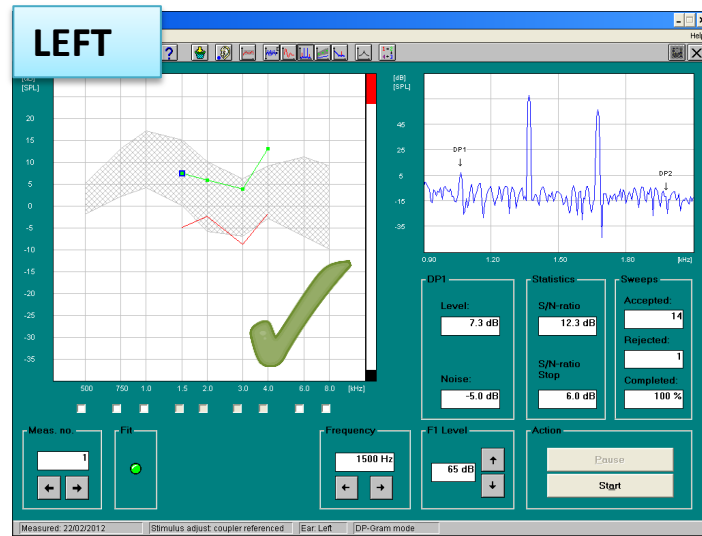
Auditory Neuropathy Spectrum Disorder (ANSD)

Tests arranged:

1. Auditory Brainstem Response (ABR)
2. Otoacoustic Emissions (OAEs)

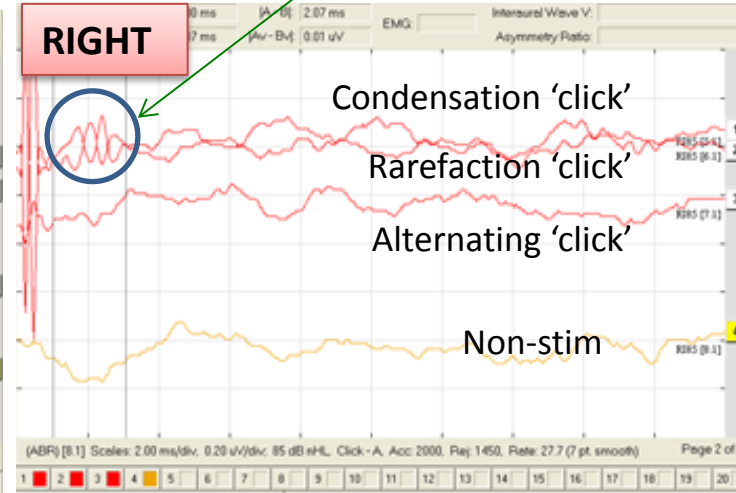
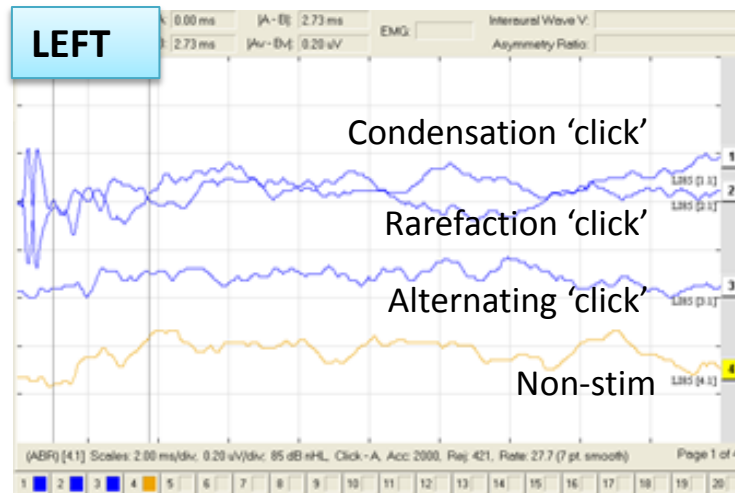


Otoacoustic Emissions (OAEs)



Auditory Brainstem Response (ABR)

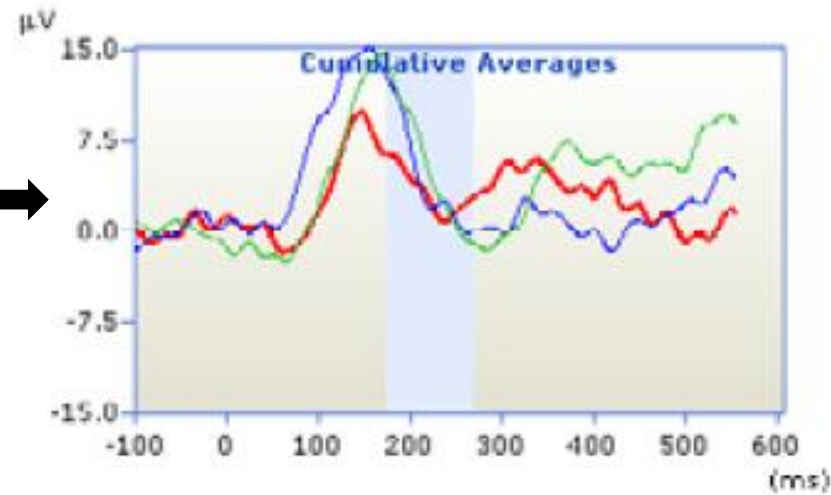
Cochlear Microphonic (CM)



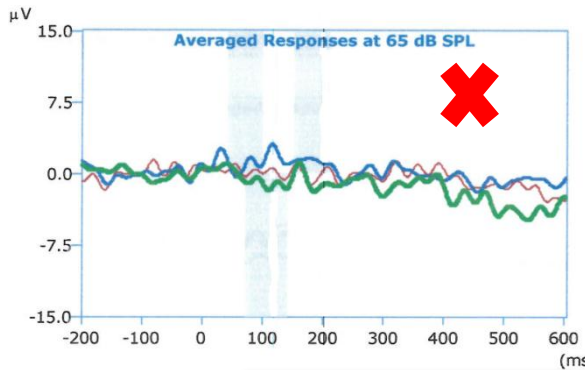
ABR confirms ANSD

Cortical Auditory Evoked Potentials (CAEPs)

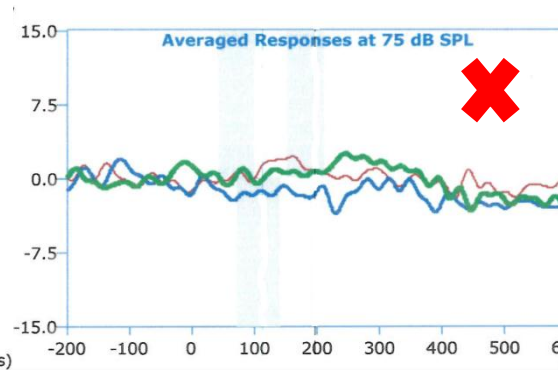
Expected to see something like this



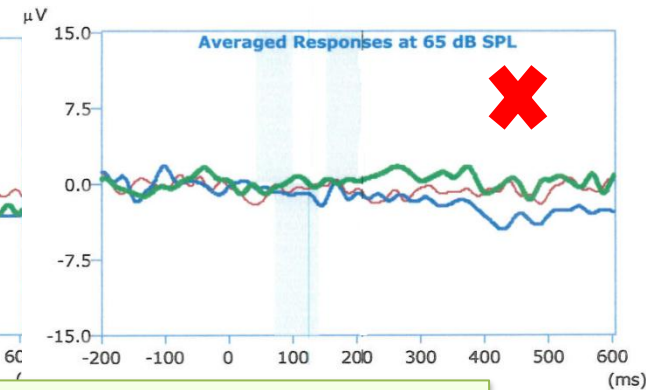
Unaided (65 dB)



Unaided (75 dB)



Aided (65 dB)



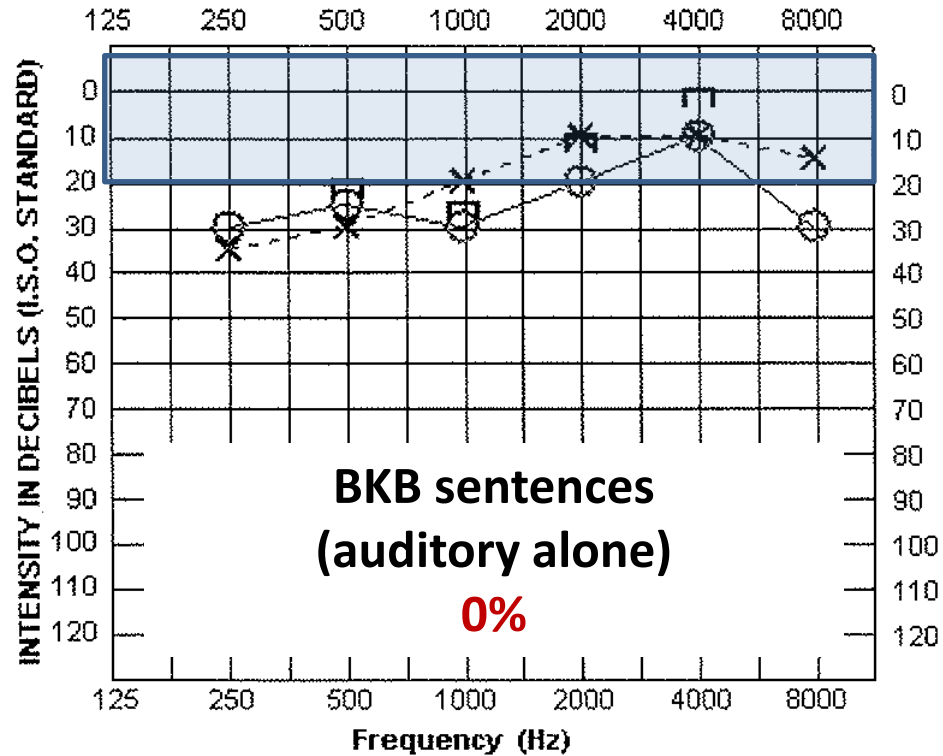
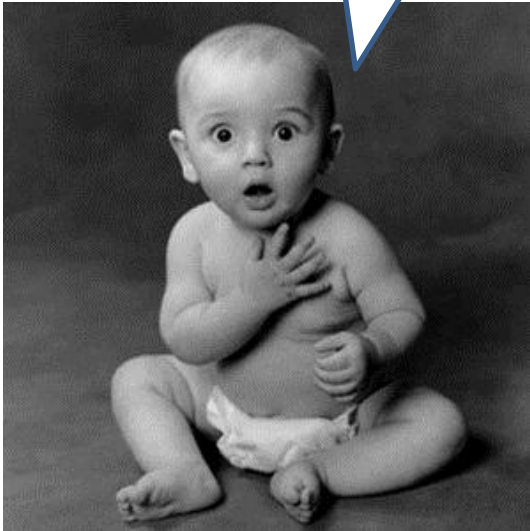
Questions:

1. What would you recommend to help this patient who is having significant problems with both vision and hearing?

Would you consider a cochlear implant?



An implant!
But it's only a mild
hearing loss!!!



AIR: LEFT X RIGHT O
BONE: UNMASKED □ MASKED RT [MASKED LT]
SOUNDFIELD THRESHOLDS □

A mild hearing loss
on an audiogram

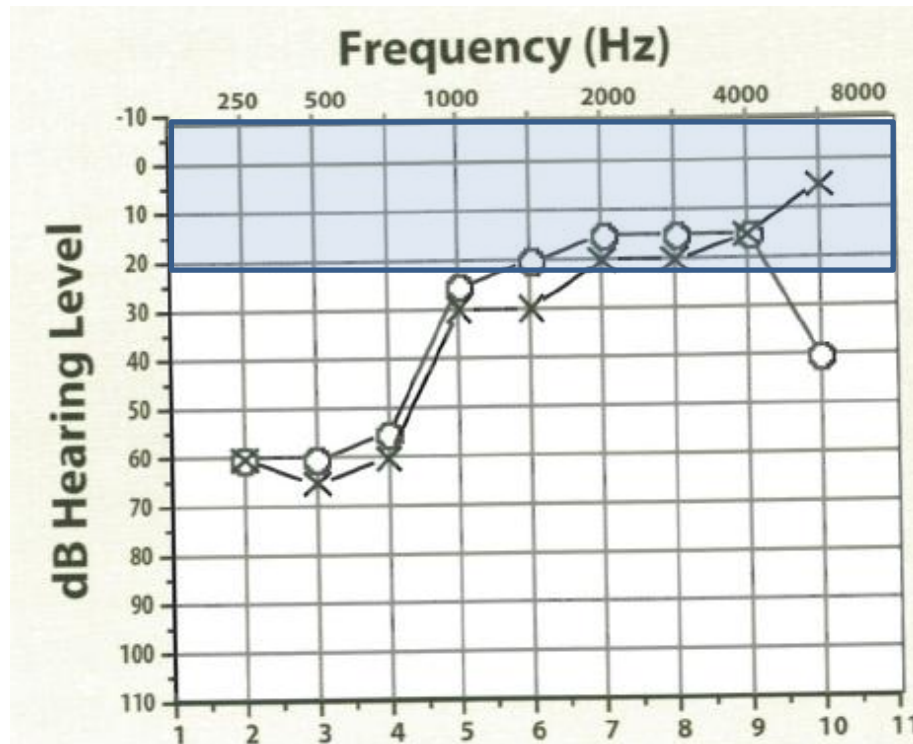


A **profound** hearing loss in
day-to-day communication

Previously reported case study

- Leber's Optic Neuropathy

Berlin et al. (2008)



	Speech discrimination scores
Pre-implant	8% & 0% (in quiet)



Patient goals

1. To be able to hear & communicate better with people
2. Improve relationships with friends & family
3. To be able to “engage” at school/college
4. To have a better future



Counselling



Worst case scenario

Doesn't like the sound from the implant & ends up not using it



- **Has lost her residual hearing in one ear.**
 - Speech may sound distorted but at least she would be aware of sounds on both sides
 - Bilateral hearing is even more important when there is poor vision
- Day-to-day life is already very stressful & if an implant “failed” it may cause further **distress**

Activity:

We asked her to block one ear with an earplug for one day & then do the same for the other ear

Our questions:

1. Did she feel disoriented when she couldn't hear as much in one ear?
2. Was there any difference between the ears?



Counselling

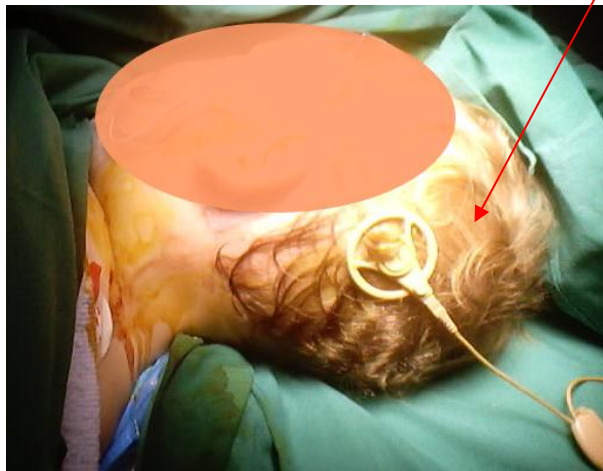
- Blocking the ear was a strange experience & did make the patient think about whether she would manage if an implant didn't work & she lost the hearing in one ear.
- She felt she heard better with the right ear open so if we implanted she would prefer a left implant
- Day-to-day life and education was becoming increasingly difficult so she felt she had to do something
- We felt comfortable that she had considered all the issues, understood it could take months to start to get used to the signal, & understood that good outcomes couldn't be guaranteed



Cochlear Implants & ANSD

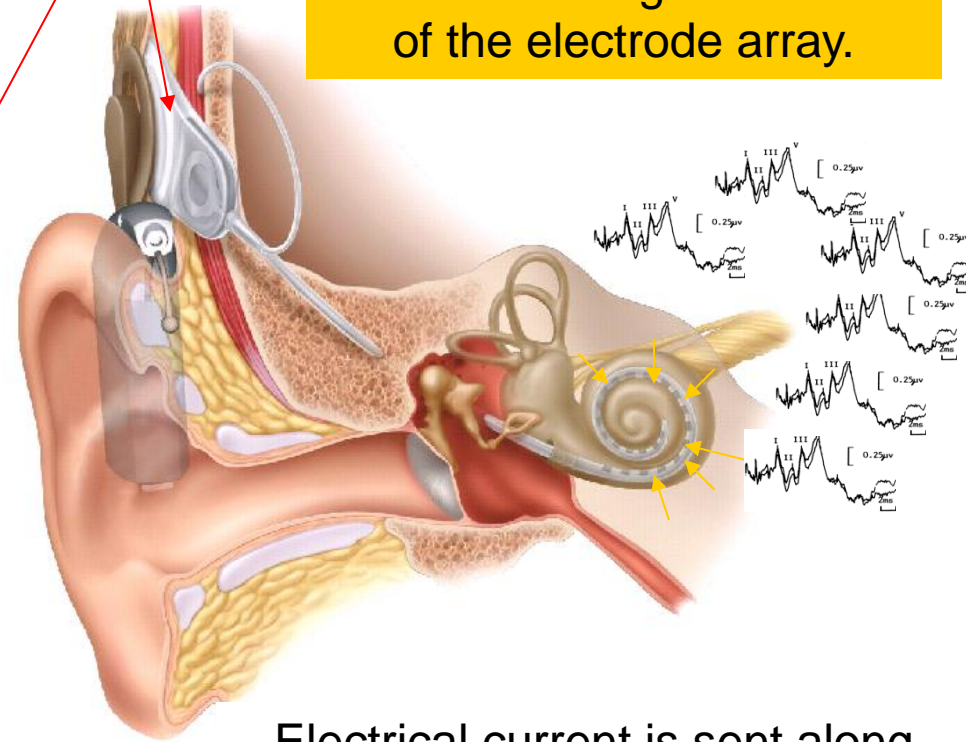


The cochlear implant is tested by the Biomedical Engineer or Audiologist as the implant site is being sewn up.



The external coil is placed over the device.

Neural survival can be evaluated along 19 sections of the electrode array.

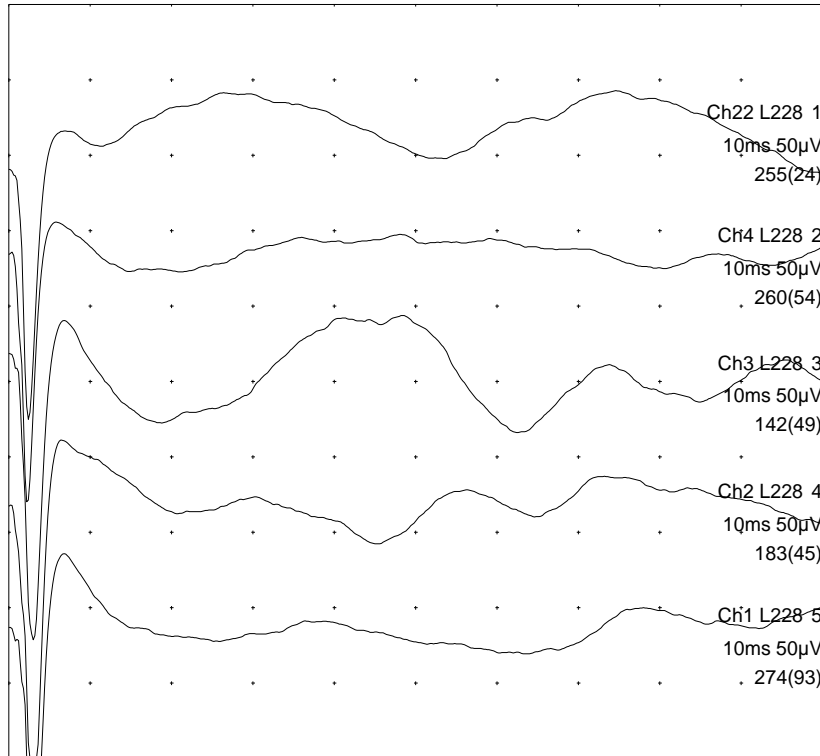


Electrical current is sent along each electrode to see how the hearing nerve & brainstem responds to the cochlear implant.

Intra-operative test results

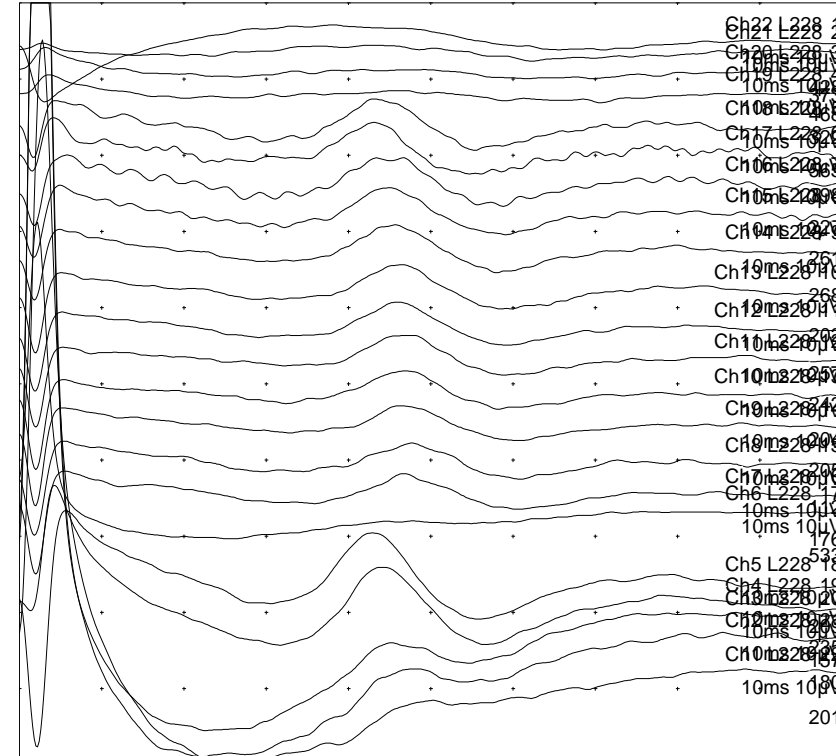
Standard pulse width

MP1+2 Ch22, 4-1



Wider pulse width

Response MP1+2



Did we make the right decision?

After 1 week:

- Can hear most of 1:1 conversation but not in groups.
- Can finally hear everything her best friend says for the first time.
- Problems hearing people with accents

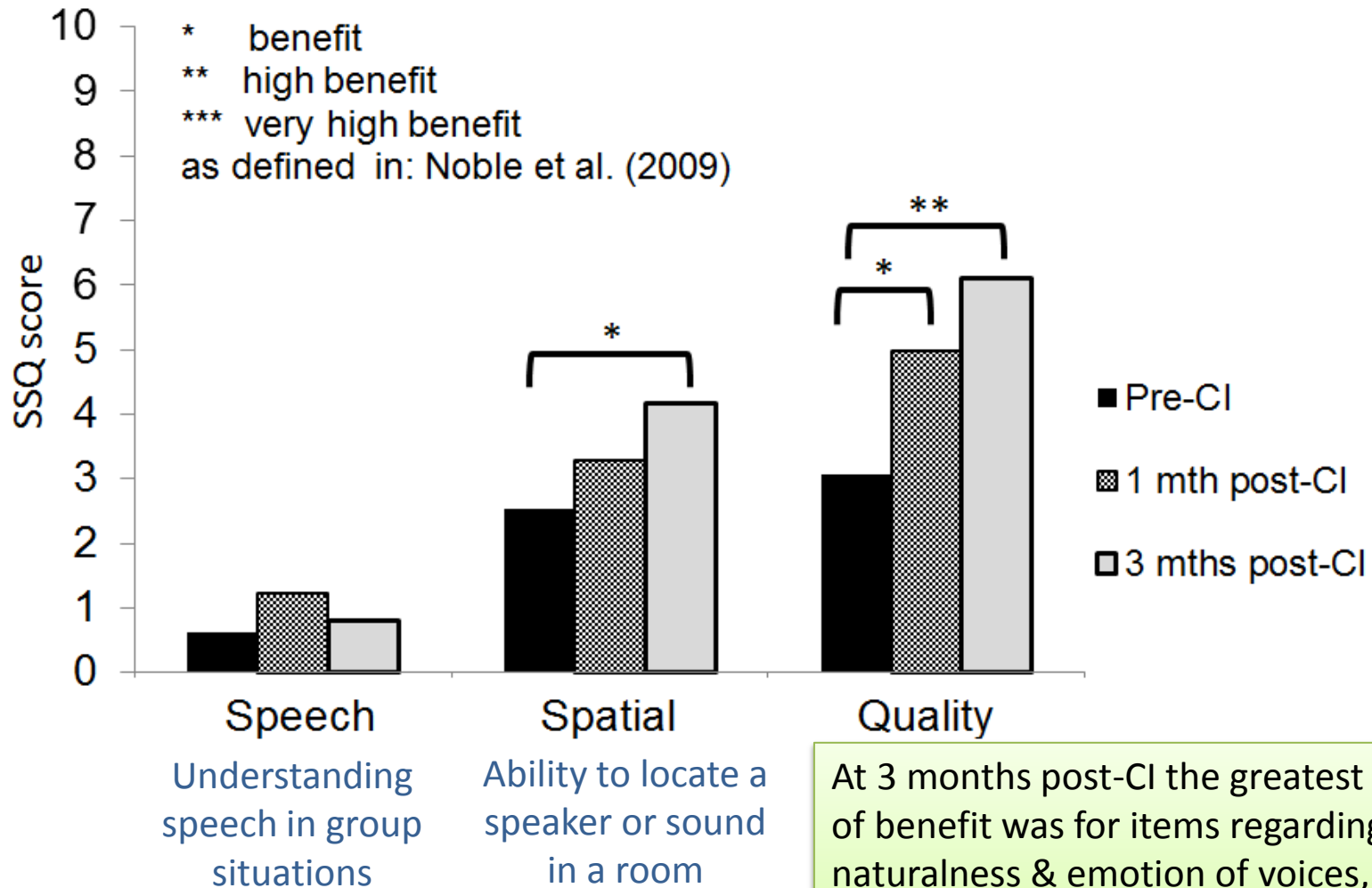


We noticed a change in confidence during follow-up appointments

Speech perception tests	Pre-CI		3mth Post-CI
	R+L	L	
CUNY sentences	52%	14%	
CVC words	22%	4%	

Changes in perceived abilities (SSQ)

Speech, Spatial, and Qualities of Hearing" (SSQ) questionnaire



At 3 months post-CI the greatest amount of benefit was for items regarding the naturalness & emotion of voices, and the effort to understand speech.

Can I have another one?!



Scheduled to have the other ear implanted later this year

Key messages:

1. Don't treat the audiogram
2. Cochlear implantation can be considered even in patients with a lot of residual hearing (particularly those with ANSD)