A Case Study in Unilateral Hearing Loss: What Phoebe has Taught Me

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Article submitted to BSA Audacity magazine for publication in Sept18.

Phoebe was born in September 2007 and was referred from her new-born hearing screen. An audiological assessment using auditory brainstem responses (ABR) suggested normal hearing in the right ear and a moderate to severe sensori-neural hearing loss (SNHL) in her left ear.

There was no family history of permanent hearing loss, and her parents requested an audiological opinion on possible cause of the unilateral hearing loss (UHL) and options for management of the hearing on the left side. The audiological physician explained that Phoebe's UHL did not fulfil the NHS criteria for hearing aid provision in the left ear. The physician also suggested that anecdotal evidence indicated that hearing aid use for UHL was not found to be beneficial by many paediatric users (though this was based on detection of hearing loss at school-age rather than newborn screening). She believed that Phoebe's speech progress was likely to be typical, as she had normal hearing in the right ear.

However, Phoebe's parents requested some further opinions on what to do about the UHL. This was because they noticed that Phoebe was less responsive to sounds than her siblings. For example, Phoebe sometimes startled when people came into view as if she hasn't heard them approach; and she could not localise to sound. Phoebe's parents' met Jacqueline Stokes, an auditory verbal therapist, who recommended that her parents consider amplification, if there was aidable hearing in the left ear.

I first saw Phoebe at 9 months of age, to assess if there was usable hearing in the left ear. This was completed using masked VRA with inserts and bone conduction, and results showed hearing that may benefit from amplification in the left side (see Figure 1, below).

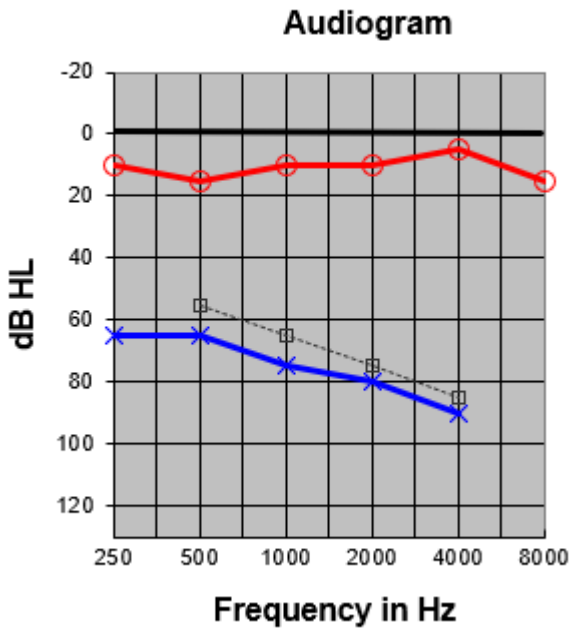


Figure 1. Phoebe doing masked VRA with inserts at 10 months and her audiometry results

Investigations into the cause of hearing loss were completed. An MRI showed an enlarged vestibular aqueduct (EVA) on the left side, and the right side was normal. This raised the possibility that the hearing on the left side might fluctuate, and possibly deteriorate. She did not have Pendreds Syndrome.

Phoebe was fitted with a behind-the-ear (BTE) hearing aid on a shell ear mould at 10 months of age. While we could verify the hearing aid output using real ear measurement, it was difficult to know if Phoebe was getting benefit from the amplification. I monitored her progress closely. Phoebe tolerated the aid well and did not attempt to remove it or show discomfort with raised sound levels.

Phoebe started to develop vocalisations with babble consonants but had no words at one year of age. Phoebe spontaneously developed some signs at home and dropped the sign once she developed the spoken word.

Of interest, on one occasion when she was 18 months old she took her hearing aid off and gave it to her mother. Her mother checked the aid and found that the battery was dead. This was the first

clear positive indication that Phoebe was benefiting from her amplification. Her family also noted that she was more responsive to sounds.

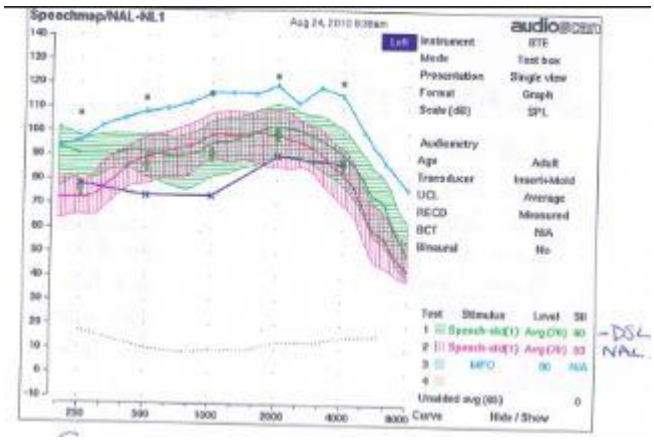
At 20 months of age, Phoebe had a vocabulary of around 50 words that were intelligible to her family. She was understanding simple instructions at home from both the right and left sides, however she could not always localise the direction that somebody was talking from.

Phoebe was seen by a speech therapist, and concerns regarding muscle tone around the mouth, and expressive and receptive speech delays were identified. Ling sounds (mm, oo, ar, ee, sh, ss) were presented with a low voice level at close range with Phoebe, with her repeating the sounds back from the right and left sides. Speech discrimination testing was assessed at 45 and 50 dB A using familiar words and Phoebe could identify these items presented from either side.

At 2 years 2 months of age, Phoebe was wearing her hearing aid through all waking hours, and her speech clarity was improving. She had an ear infection in her left ear at around 3 years of age and was not able to wear her hearing aid for a week. Phoebe specifically asked on Sunday night “Please can I have my hearing aid back now?”.

As Phoebe shows some startle responses for sudden sounds, like coughing or sneezing, when wearing her hearing aid, we adjusted the hearing aid fitting to match NAL/NL2 targets rather than using the more typical DSL prescription. The overall loudness density of DSL is greater than amplification with the NAL prescription. When we put the hearing aid back on with the new NAL prescription setting, she said “my hearing aid is not working now”. A hearing aid check found it to be working well. I believe this is because the overall perceptual level was less than her hearing of audible speech in her normal hearing ear, and therefore she was not aware that the hearing aid was on. I interpreted this as a functional demonstration of the Stenger principle.

Figure 2 NAL and DSL Curves for Phoebe’s Hearing Loss

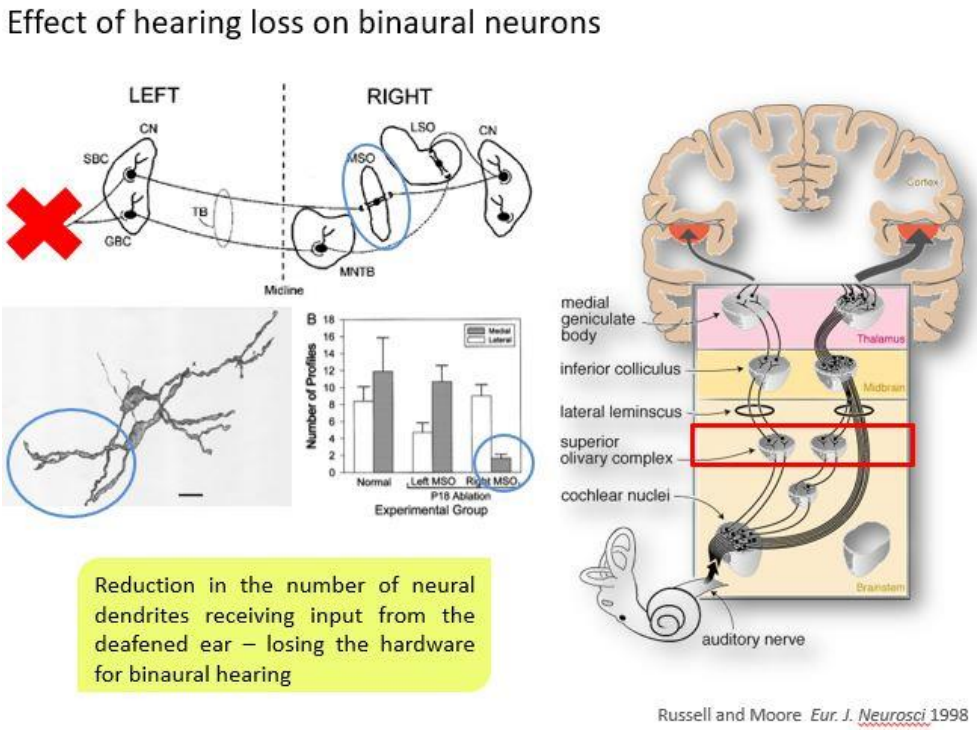


The Stenger effect is a known perceptual effect where by a sound is delivered to both ears simultaneously and is heard only in the ear with the higher intensity signal. Thus, it is interesting to note, that for Phoebe’s hearing configuration, the overall perceptual level for a NAL fitting is slightly less than for normal hearing (that she has in the right ear), while DSL has a slightly higher perceptual level than normal hearing.

A second possible explanation for Phoebe’s jumpiness to loud sounds was also discovered. Was her jumpiness to sudden sounds less with the NAL hearing aid fitting? Her mother clarified that Phoebe was jumpy to sudden sounds both with and without her hearing aid on. When I discussed this event

with David McAlpine at the Ear Institute, he directed me to a paper describing this effect (McAlpine et al, 1997). The paper described the impact of UHLs and the lack of suppression at the level of the inferior colliculus to the contralateral (normal hearing) ear. This was new learning, as what I had always considered to be a “normal-hearing ear,” in fact had reduced suppression, and sensitivity to sudden high levels of sound. On reviewing file notes of my other children with UHL, I found that this was consistently reported by many of their parents.

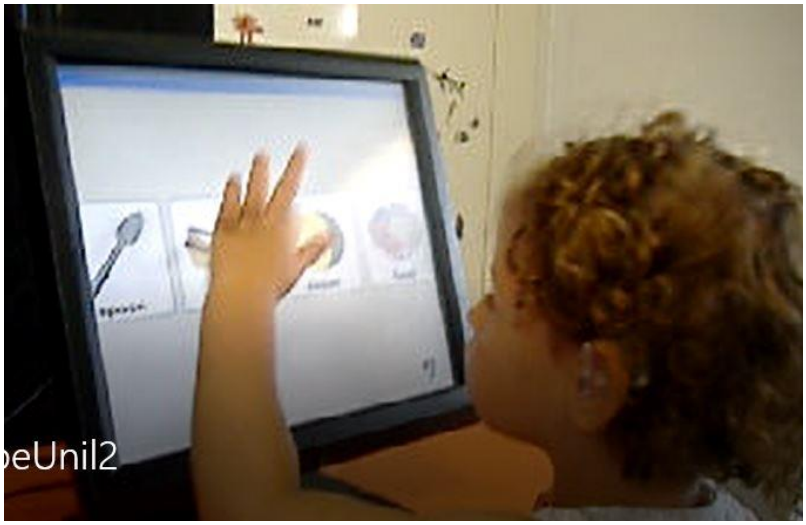
Figure 3 showing the effect of hearing loss on binaural neurons in deafened ferrets



(With thanks to David McAlpine for permission to use this figure)

At 3 years of age, speech discrimination testing using computerised presentation of four words (CCT) on a touchscreen was completed. When tested with and without her hearing aid, Phoebe scored 82% in both conditions. To access more high frequency sounds, Phoebe was fit with a new hearing aid that had frequency lowering technology. Phoebe was fit with DSL, as this seemed to be less associated with startling to loud sounds. With her new hearing aids, she scored 90%.

Figure 4. Phoebe doing speech testing at 3 years with her left hearing aid on



At 3 ½ years Phoebe started in Nursery and chose to use her hearing aid throughout the school day. She liked it particularly in group settings, and during phonics. Her hearing in her left ear remained stable and unchanged.

Phoebe was able to provide clear reports of hearing difficulties with following conversation in noisy environments (e.g. family group or restaurant). Phoebe began to report on her localisation abilities. She reported that sometimes she could localise to sounds with the hearing aid, but in other situations she could not. Phoebe shared a wonderful experience from school saying, "Do you know, there is a girl in my school who has two hearing aids. How lucky is she?"

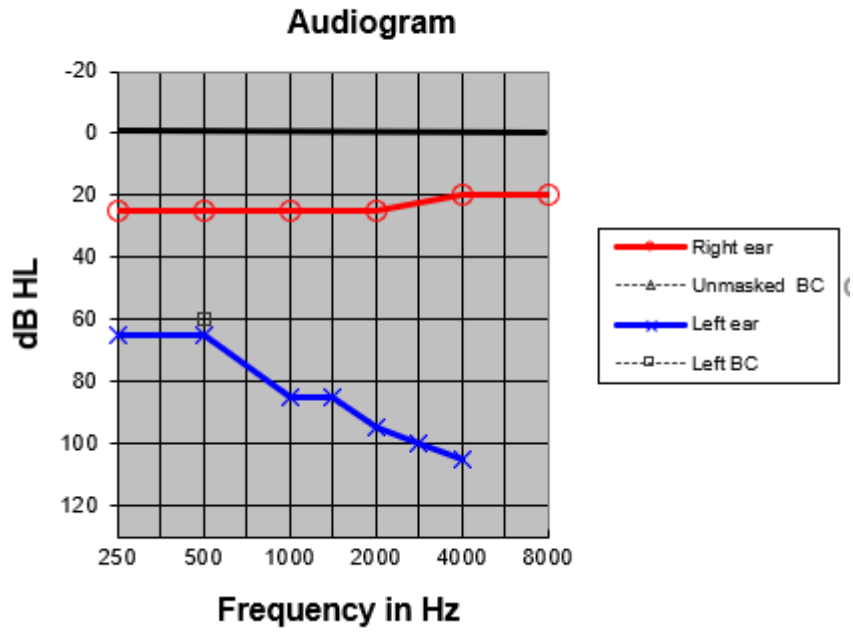
By 4 years of age, Phoebe could complete speech discrimination testing with open-set speech tests with masking noise in her good ear, to assess her speech discrimination for each ear separately. On open-set speech test, Phoebe scored 100% correct at 65 dB and 80% correct at 50 dB presentation level listening through her left hearing aid.

At 6 years of age, Phoebe was changed from a full shell ear mould to a receiver-in-the-canal (RIC) hearing aid. This was for improved comfort and for better access to high frequency speech sounds. In addition, Phoebe used a radio aid receiver in her left ear, for easier access to the teacher's voice in the classroom. Phoebe advocated well for herself at school; she had a buddy system and was insightful about the best position for her in the classroom.

At the age of 8 ½ years, Phoebe noticed a change in her hearing on the left side. This was of concern because Phoebe was known to have enlarged vestibular aqueduct (EVA) with an increased possibility of fluctuations in cochlear hearing. According to the literature, 30% of enlarged vestibular aqueduct condition had evidence of endolymphatic hydrops. In addition, research has reported that where the MRI shows a unilateral enlarged vestibular aqueduct, there can be susceptibility to bilateral deterioration in hearing.

A hearing assessment revealed that her hearing had deteriorated on the left side by between 15 - 20 dB across all frequencies. Because of this decrease, Phoebe's hearing aid was changed to a superpower receiver in the left ear. Also, of particular concern, were her hearing levels in her right ear. They were now fluctuating, and were just outside the normal hearing range (Figure 5.)

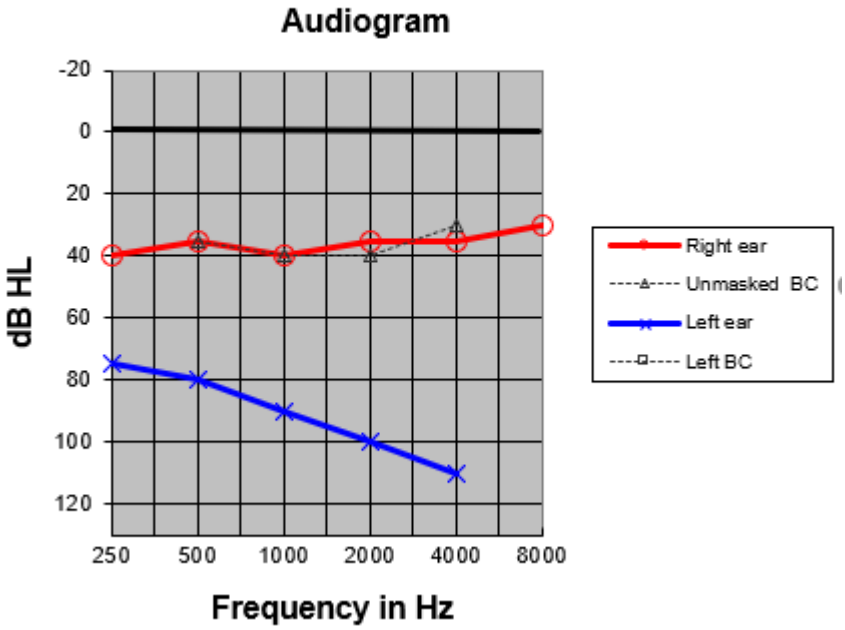
Figure 5. Hearing levels deteriorated in both the left and right ears



At 9 years, 2 months of age Phoebe had an episode that she described as “blurry hearing.” This occurred during in a lesson in the school day and improved over the course of the week. In her normal cheery way, Phoebe described that “No one could tell me off for five days.”

This appeared to be an early indication of cochlear hydrops, which gave rise to a further deterioration in hearing in both ears (Figure 6).

Figure 6. Audiometric thresholds at 9 years



Phoebe was seen by her audiological physician, and her family were advised that they should reduce sugar and salt in her diet to reduce susceptibility to hydrops attacks. To address the changes in hearing in her right ear, Phoebe was fitted with an open fit hearing aid with an integrated radio receiver.

Given the deterioration in her left ear, and the uncertainty regarding the stability of hearing in her right ear, her audiological physician recommended the family consider a cochlear implant in the left ear. As part of the family's consideration, Phoebe attended a Cochlear Implant Information day at the Ear Foundation, spoke to the Cochlear Implanted Children's Support Group (CICS), and met other children of her age with cochlear implants. Initially, she was reluctant to think about a cochlear implant. She was seen by Professor Shak Saeed at UCL who recommended a left sided cochlear implant. While the current NICE guidelines from the NHS do not include cochlear implant for UHL, the fluctuating hearing in her right ear made this a more compelling consideration for Phoebe and her family.

At 9 years, 4 months of age, Phoebe was seen by a neurologist. This was because she had had a migraine, dizzy spells and tinnitus in both ears over the Christmas holidays. She described the tinnitus in her right ear as sounding like a lamb sound "baa," while the left ear had intermittent high-pitched tinnitus. Unfortunately, the left hearing aid no longer provided any benefit for Phoebe. She continued to use her right hearing aid, and this was helpful in the classroom. While Phoebe was able to maintain her progress in school, she struggled socially. She couldn't interact with friends at lunch or in the playground and was very tired by the end of every school day.

In April 2017 at the age of 9 and 6 months of age, Phoebe received a cochlear implant in her left ear. Unfortunately, Phoebe had vertigo (a "vomit-o-thon") for about 10 days following the cochlear implant surgery. This was a recognised potential complication from cochlear implant surgery in EVA.

Although some studies have indicated significant variation in tolerance, and speed of adoption of a cochlear implant, Phoebe reported that it sounded like normal speech after only one day. Even after only a few weeks of use she said the implant was "Worth it 100 times over!" She also stated that her

hearing was “so much better than previously with her hearing aid.” Phoebe was actively involved in the programming of her CI device, and reported that the clarity of speech on her left side was “great”. While Phoebe is not using a radio aid or Bluetooth streamer, she is thriving in school including in dictation. She reports that sudden noises are still sometimes too loud and that the worst listening situations for her are large family dinners. Phoebe reported that she listens to music through her Implant, without any distortion.

Phoebe finds her one-piece speech processor less visible than her previous hearing aid because the colour blends with her hair. She also feels it is much more comfortable than her hearing aid, as there is no piece over her ear.

Figure 7. Phoebe with her cochlear implant

Phoebe currently has a moderate low-frequency sensorineural hearing loss in her right ear. She has had a period of normal hearing over about 2 months which then became moderately impaired again. She uses a hearing aid on the right side when needed but uses her cochlear implant on her left ear through all waking hours.

What have I learned from Phoebe over the last 10 years?

1. The importance of optimising hearing where there is potential benefit from amplification. Phoebe has taught me to be creative in finding ways to functionally verify hearing of speech

through hearing aids. A unilateral hearing aid can be tolerated well, even loved, if introduced from under a year.

2. Her family have shown me the importance of managing hearing, both for neural stimulation and for improving functional hearing on a day-to-day and week-by-week basis. Her father once asked the group of students at an Ear Institute course on unilateral hearing loss "If your child was blind in one eye and you could do something about it, you would, wouldn't you? Why is that any different to hearing loss in one ear?"
3. Phoebe has taught me how resilient young people are and how they can drive the process when necessary. If we hadn't aided Phoebe's left ear all those years ago we wouldn't have had the same opportunity for benefit from cochlear implantation when the hearing deteriorated in both sides. About 8 months post-op, Phoebe said "No one will ever understand the difference this implant has made to my life."
4. I understand a bit more about the perceptual loudness from different prescriptions (NAL and DSL) for hearing aid fittings and how the Stenger effect that we use for clinical testing impacts on everyday functional listening.
5. I've learned that I need to be following the evolving science in auditory perception. For example, that unilateral hearing loss has reduced suppression at the level of the inferior colliculus, that EVA on one side can give rise to fluctuating hearing loss in both ears, and that hydrops may co-occur in about 30% of cases with EVA.

Being a paediatric audiologist always gives opportunities for us to learn and I'm so grateful for having had this chance to know Phoebe and her family in her extraordinary journey so far.

Reference

McAlpine D, Russell M, Mossop J, Moore D. (1997) Response properties of neurons in the inferior colliculus of the monaurally deafened ferret to acoustic stimulation of the intact ear. J Neurophysiol 78:767-779.

JEMPhoebe4/18.06.18