So, baby, how does it sound? Cortical assessment of infants with hearing aids

By Harvey Dillon

Let’s start with a double-barreled question: Why is NAL messing with electrophysiologic potentials, and why do you think they should be part of fitting hearing aids to an infant?

Six or seven years ago, Denis Byrne and I were discussing the likely impact of universal newborn hearing screening on hearing aid fitting practice. We reasoned that its advent would create a strong need for methods to verify the suitability of a hearing aid fitting for an infant. After all, one can’t ask infants the inevitable hearing aid question: “How does it sound?”

Shortly after, Suzanne Purdy joined NAL and established an electrophysiology laboratory to tackle this question. The basic concept of using electrophysiologic responses to verify a hearing aid fitting has a long history,1-4 but until now there have not been any practical methods to do so. Suzanne’s experience with evoked cortical responses led her to believe that they had the potential to provide appropriate verification of the fitting.

That makes the next question easy. Why measure cortical responses rather than the more familiar auditory brainstem response (ABR) or auditory steady-state response (ASSR)?

Evoked cortical responses have several characteristics that make them suitable for evaluating perception with a hearing aid.

First, the stimuli can be longer than the brief clicks or tone pips that are needed to measure an ABR. This means the hearing aid has time to react to the sound (e.g., for a compressor to operate) in a manner similar to what it does for an ongoing speech signal.

Second, because the response arises from the auditory cortex, just under the scalp, the response amplitude is much larger (around 5 to 10 microvolts) than the amplitude of an ABR or ASSR signal that originates deep within the brainstem, so less stimulus repetition is needed.

Third, and most importantly, they originate from neurons at or near the end of the auditory chain, so they are affected by the hearing aid response and all parts of the auditory system, rather than just sensing the signal part way through the auditory system.5 Consequently, the responses are more likely than earlier electrophysiologic responses to correlate with perception.

Fourth, evoked cortical responses can (and indeed must) be measured when the baby is awake, and as babies get older, it gets more and more convenient to have a test that works when they are awake rather than when they are asleep. The absence of a cortical response when a person is asleep reflects the close association between the response and perception itself.

Finally, cortical responses can be measured for speech sounds, which, in the end, are what we are most directly interested in.
Before you tell me how you do the test on babies wearing hearing aids, can you tell me what the response looks like when you do an evoked cortical response test on people with normal hearing?

The response we are using is called an obligatory response. That means that the response waveform appears, irrespective of whether the person receiving the sound attends to the sound or completely ignores it. For an adult, the response waveform is characterized by a small positive peak (called P1) about 50 ms after stimulus onset, a large negative peak (N1) about 100 ms after stimulus onset, and a second large positive peak (no prizes for guessing it’s called P2) about 200 ms after stimulus onset.

The shape is very different, and more variable, for infants, often comprising just a single broad peak around 200 ms after stimulus onset. The shape changes as the auditory cortex matures, right through the teenage years up to early adulthood.

You have talked about the response shape, but what is the test stimulus?

It can be anything, with both long tone bursts and short speech sounds being used in the past. We have chosen to devise a test based on three speech sounds: the consonants /m/, /g/, and /t/. We chose these three because their frequency spectra have a low-, mid-, and high-frequency emphasis, respectively, so we figured the cortical responses could tell us something about the infant’s perception of low-, mid-, and high-frequency speech sounds.

Does someone with normal hearing show the same response no matter what speech sound is presented?

Good question. Other researchers have shown that, averaged across groups of people, different speech sounds lead to slightly different latencies. As we wanted to devise a clinical test that would work with an individual, we looked around for a way to test if, for a particular person, the responses to two or more sounds are different.

We devised a way to apply Multivariate Analysis of Variance (MANOVA) to test whether or not the wave shape as a whole changes as we change the stimulus. Essentially, we break up each individual response into a series of time bins. If the response to one sound really is different from the response to another, then for at least one time bin the mean value for one sound will be different from the mean value for the other sound by an amount bigger than that expected from chance, given the variation observed among the successive responses to repeated sounds. This would be a bit like the t-test that you studied in statistics.

MANOVA actually does better than this because it simultaneously takes into account all the time bins, even if some of the changes are positive and some negative. Pleasingly, 19 out of 20 babies with normal hearing showed a statistically significant response to /g/ versus /m/ and to /t/ versus /m/. About half the babies also showed a significantly different response to /t/ versus /g/.
You said you chose three speech sounds because of their frequency spectra. What about other speech sounds?

We have subsequently experimented with further sounds, both consonants and vowels, in infants and adults with normal hearing. Although all sounds tested reliably produced cortical responses, only certain pairs reliably produce different wave shapes, so we have stuck with our original choice of /m/, /g/, and /t/ for the clinical test.

Earlier you implied that infants have a less easily identified response shape than adults. How can an audiologist tell if a response is actually present?

Fortunately, the same statistical procedure I told you about earlier (MANOVA) can also be used to determine if a response is present. In fact, our very recent research (using adult subjects) has shown that the MANOVA technique is more accurate than expert clinicians at detecting a cortical response.

The technique does not make any a priori assumptions about the shape of the response, so it should be just as accurate for the less predictable infant responses as it is for adult responses. The same is probably not true for human observers, who are partly reliant on peaks or troughs being present at particular times.

How does all this lead to a verification procedure for the hearing aid fitting?

We reasoned that if a baby wearing aids shows a cortical response when these three speech sounds are presented at typical speech levels (55 to 75 dB SPL) in the sound field, and if the responses are reliably different for, say, /m/ versus /t/, then it seems likely that the baby is getting sufficient information from the amplified speech to learn to make at least some use of it.

If you don’t see a cortical response, does this mean the child didn’t hear the sound?

We think so. In the experiment I just mentioned, both MANOVA and the experts were very good at detecting a response when it was 20 dB above behavioral threshold. MANOVA continued to give almost perfect performance when the sound was only 10 dB above behavioral threshold. Even responses for sounds right at behavioral threshold were detected more often than not, so if no response is detected, our conclusion (at least for adults) is that the sound is either inaudible or is at most 10 dB above threshold. Neither inaudibility nor a sensation level so low that there is no cortical response is likely to be conducive to good speech perception.

We need more research on the relationship between cortical threshold and behavioral threshold for infants with different degrees of loss, but it was shown 40 years ago that cortical responses can be measured at or very close to threshold for older children with hearing impairment.

No exceptions?
Actually, there are. The cortical response of some children with cerebral palsy, for example, will have a very high noise level because of the frequent and strong muscular activity around the head. Consequently, a cortical response may not be detected, even if one is, in fact, present, but buried in the noise.

11 If two responses are different, does this mean the infant can tell the sounds apart?

We have no direct evidence of that, but if the response shapes are different for two speech sounds, then *something* different is happening inside the baby's brain for one sound than for the other. It seems a reasonable presumption that the baby will be able to learn to use these differences to discriminate the sounds.

Note that we don't expect the response shapes to look normal right from the time the hearing aids are fitted. The latency of cortical responses is affected by the child's previous auditory experience, and lack thereof. Specifically, latency of the major positive peak is longer than normal for children who have had inadequate exposure to sound because of a congenital hearing loss, but then shortens following access to sound. For a baby with a severe loss receiving his or her first hearing aids, previous auditory experience is likely to be very limited.

12 What actual evidence is there that children showing cortical responses make better use of their hearing aids than children with no responses?

The most direct evidence comes from Rance et al., who showed that for older children with auditory neuropathy (which in this article will include dys-synchrony), the presence of cortical responses was a nearly perfect indicator of the child's ability to understand speech through the hearing aid. Obtaining further evidence for babies with varying degrees of sensorineural hearing loss is a current research area of ours. Maryanne Golding is analyzing the results of 40 infants who have been assessed both by aided cortical assessment and by a functional assessment method that enlists the parent to examine how well the child responds to sounds in their usual environment.

13 What do you do if a response is not present for any of the speech sounds?

The worrying implication is that if the child is not hearing these speech sounds at typical speech levels, then the child is most unlikely to have adequate speech perception.

The first thing to do would be to ensure that everything done up to that point is error free. This includes the hearing threshold measurement (in the case of infants, presumably based on ASSR or toneburst ABR), estimation of behavioral thresholds from these measurements, the hearing aid prescription process (preferably using either NAL-NL1 or DSL[i/o]), and adjustment of the hearing aids to achieve the prescription. I probably should point out that aided cortical evaluation is not a replacement for verification of gain and output using probe-microphone measures, preferably accomplished by measurement of the real-ear-to-coupler difference (RECD). Rather, the two measures are complementary.

Even if no error can be discovered, the hearing aid gain should be increased and the measurement repeated, as something is wrong.
if there are no responses present. If it is simply impossible to obtain responses, further investigation of the child’s hearing loss is called for, and the continued absence of response is an important consideration in deciding if a cochlear implant is warranted.

I am assuming that the response waveform is not excessively noisy, and that the child was awake when the measurement was done. No valid conclusions can be drawn from measurements that indicate excessive noise or from sleeping children. If there is a response to only one or two of the sounds, consideration should be given to increasing the gain of the hearing aid in the region where the missing speech sounds have their primary emphasis.

14 Do the responses actually change if the hearing aid is adjusted?

Yes. We know this because we have investigated in a group of older children, again using the MANOVA technique, whether or not the presence and differentiation of the responses change as the gain-frequency response of the hearing aid is altered. As we introduce a low-boost, high-cut, or vice versa, the response shape significantly changes more often than not. These results are currently being written up by Maryanne Golding at NAL.

15 How long does the testing take?

Currently it takes a couple of hours to perform tests, unaided and aided, on three speech sounds, at two input levels, and to have the associated discussions with the parent. As part of this testing we aim for a more comprehensive picture of what the baby can hear: We increase the level to 75 dB SPL for those sounds where there is no response at 65 dB SPL, and we decrease the level to 55 dB SPL when there is a response at 65 dB SPL.

We are currently incorporating the MANOVA technique directly into some test equipment, and we think that we will be able to reduce total time to less than an hour.

16 What else do I need to know about the testing?

Test one ear (and hearing aid) at a time by simply turning off the hearing aid in the other ear. Position the infant or loudspeaker so that the loudspeaker is either directly ahead or a little to the side of the head being tested. Keep the infant in an entertained and awake, but not noisy, state. Soft toys are good. Usually the infant will be sitting on the lap of a parent and everything (the parent, child, loudspeaker, clinician, and equipment) can be in the same test room.

Remember that, although the test is very good at telling you when the child cannot hear the speech sounds and can provide reassuring information that different sounds are being processed differently by the brain, it cannot tell you when a hearing aid is overamplifying sounds. Fortunately, the combination of WDRC (which we recommend for all children) and the maximum output prescriptions built into both NAL-NL1 and DSL[i/o] make us less fearful of overamplification than we were in the past.
How do parents react when they see the absence or presence of brain wave activity in response to sounds?

By the time the hearing aid fitting takes place most parents have accepted the presence of a hearing loss. If they have not, then the absence of a response when the child is unaided provides further confirmation to them that their child has a loss.

Conversely, if the parents are overly pessimistic about the future for their child, the appearance of the responses while the child is listening to typical speech sounds and wearing the hearing aid can be extremely reassuring, as it shows very directly that the child is perceiving speech. We cannot, however, make any predictions about the child’s speech-discrimination ability.

Are there any infants for whom aided cortical assessment is particularly valuable?

Definitely. The less certain you are about the hearing loss of the child, the greater the need to verify that the child is hearing a range of speech sounds when amplified. One such group of children are those with auditory neuropathy.

The state of New South Wales, where NAL is situated, has a universal newborn screening program that includes automated ABR. In the first year of operation, 15% of the children detected had symptoms consistent with auditory neuropathy (otoacoustic emissions and/or cochlear microphonic present but no ABR response). We have subsequently seen most of these children at NAL and, in several cases, behavioral responses when the child is around 1 year of age are more consistent with the earlier cortical responses than with the earlier ABR results.

A second group includes those children with severe loss for whom no thresholds could be measured at one or more frequencies. For such children, hearing aid fitting has traditionally involved a prolonged trial-and-error approach. The aided cortical evaluation allows this to be considerably speeded up. In general, we have found that the clinicians responsible for the children value the cortical results whenever there is any reason for them to doubt that the hearing has been appropriately adjusted.

Are there patients besides infants for whom the measurement of cortical responses might be useful?

The traditional application remains valid. In many parts of the world, cortical responses have long been regarded as the gold standard for measuring thresholds, using pure-tone stimuli, when non-organic loss (i.e., exaggerated loss) is suspected, because agreement between evoked thresholds and behavioral thresholds is so good.

Also, patients with multiple disabilities or other patients who are unable to give behavioral responses reliably can conveniently have their thresholds measured using evoked cortical responses. We have several ideas for how our statistical technique and some other novel ideas can be employed to facilitate cortical testing for both aided cortical assessment and threshold determination.
That all sounds great. Do you believe that in the near future aided cortical assessment will become routine for all infants fitted with hearing aids?

From the reaction of clinicians in the hearing centers around NAL who are currently sending their infants to us to have this test done, I am sure it will become standard practice for all infants with severe or profound loss, all infants with auditory neuropathy, and all infants where, for any other reason, there is uncertainty over their hearing thresholds. I think it’s likely that it will get used for all infants, as it is just so reassuring to see the responses emerge after you believe everything has been done correctly. Even more importantly, it provides clear evidence when you need to do more.

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REFERENCES