Auditory Brain Development in Children with Hearing Loss – Part Two

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Editor’s Note: This is the conclusion of a two-part article. The first part was published in the October 2016 issue.

5. The secondary auditory cortex: For sale to the highest bidder!

Although the activity observed in the primary auditory cortex was certainly interesting, the most relevant finding of the Nishimura et al. study was the activity they observed in the secondary auditory cortex (Nature. 1999; 397[6715]:116). Little to no activity was observed in the secondary auditory cortex while participants listened to running speech, but robust neural activity was observed in the area when participants observed sign language (Fig. 6). This finding was one of the most prominent early reports of cross-modal reorganization in the secondary auditory cortices of people who are born with severe to profound hearing loss and deprived of access to intelligible speech during the first few years of life.

Stated differently, in the absence of access to intelligible speech from the primary auditory cortex, the secondary auditory cortex is colonized by the visual system to aid in visual function. Numerous published reports have shown similar findings over the past 15 years, with some indicating activity in the secondary auditory cortex in response to tactile stimulation as well (Brain Res Rev. 2007;56[1]:259). The acquisition of the secondary auditory cortex by other sensory modalities likely explains why people who are born deaf without sufficient access to auditory stimuli develop exceptionally adept abilities in some areas that involve other sensory functions (e.g., peripheral vision is better in people who are born deaf without access to sound during the critical period) (Trends Cogn Sci. 2006; 10[11]:512). Since such reorganization occurs outside of the primary auditory cortex, a functional disconnection between the primary and secondary cortices was postulated (Brain Res Rev. 2007;56[1]:259).

Recent research in Dr. Andrej Kral’s laboratory investigated activity in single neurons in the secondary auditory cortex in response to cochlear implants (CIs) (J Neurosci. 2016; 36[23]:6175). They demonstrated that while some anatomical fiber tracts among cortical areas and thalamus persist in deafness and the secondary cortex preserves some auditory responsiveness, there is also an increased visual responsiveness in the area (PLoS One. 2013;8[4]:e60093). Neurons in the secondary auditory area that responded to visual stimuli did not respond to auditory stimuli, demonstrating that visual input occupied some of the auditory resources normally used by hearing.

The results of Dr. Kral’s studies (along with the research of others) suggest that when the brain does not have access to intelligible speech during the early years of life, meaningful auditory input does not coordinate activity between the primary and secondary auditory cortices. Instead, the secondary auditory cortex assists with other sensory functions such as visual processing. Additionally, auditory stimulation beyond the critical period of language development finds disordered functional connections or interactions between the primary and secondary auditory cortices, further limiting auditory learning.

4. The break-up! Starring the primary and secondary auditory cortices

At this point, a natural question to ask is, “Where does the disconnect occur when the auditory areas of the brain do not
receive early access to intelligible speech?” To answer that question, we turn to Dr. Kral’s research exploring the functions within the multiple layers of the primary auditory cortex. The auditory cortex is comprised of six layers of neurons (2-4 mm thick; Fig. 7). Afferent inputs from the thalamus arrive at the cortex at layer IV, and much of the processing within the cortex takes place at layers I-III (i.e. the supragranular layers). Layers V-VI (i.e., the infragranular layers) modulate activity in the supragranular layers, serve as the output layers of the cortex into the subcortical auditory structures, receive top-down projections from higher-order areas, and integrate higher-order information with the bottom-up stream of auditory input.

Dr. Kral measured neural responses to auditory stimulation at the different layers of the auditory cortex using microelectrodes inserted to varying depths (Cereb Cortex. 2000;10[7]:714). Because such testing is too invasive to conduct in young children, Dr. Kral completed his studies with deaf white cats. He discovered activity in layers I-IV but reduced activity in layers V-VI (Fig. 8). Among other deficits, reduced infragranular layer activity interferes with the integration of bottom-up and top-down information streams. As a result, Dr. Kral and his coauthors concluded that a functional decoupling between the primary and secondary auditory cortices had occurred, particularly from the top-down information stream.

This “break-up” between the primary and secondary cortices has significant functional implications for auditory and spoken language. When auditory signals are not efficiently and effectively transmitted from the primary to secondary auditory cortex, the secondary cortex cannot share spoken language and other meaningful sounds with the rest of the brain. This lack of distribution of auditory stimulation to the secondary auditory cortex and then to the rest of the brain explains why a teenager who was born deaf and never had access to meaningful auditory stimulation would not be able to develop, support, sustain, or lay a foundation for a rich and robust auditory system that has been exposed to a rich and robust auditory system that has been exposed to meaningful auditory stimulation. Indeed, visual stimulation elicits responses in the secondary cortex, but it does not promote the development of functional synaptic connections between the primary and secondary auditory cortices, which are required for meaningful auditory stimulation to be disseminated in a neural network across the brain.

3. Kids and kittens

In deaf white kittens, Dr. Kral showed that the loss of infragranular activity developed remarkably early (Cereb Cortex. 2002;12[8]:797; Brain. 2013;136[Pt 1]: 180-93). In other words, the critical period of auditory brain development was not provided during these first few months, during the time when synaptic development happens, development of the auditory function is severely compromised. However, if these deaf kittens were provided with a CI within this time period (Fig. 8), the microelectrode recordings made by Dr. Kral and his colleagues suggest the kittens’ auditory areas of the brain developed rather typically.

How does Dr. Kral’s research with kittens translate to kids? For that answer, let’s turn to the work of Dr. Anu Sharma, who measured the latency of the P1 component of the cortical auditory evoked potential (P1-CAEP) in children with normal hearing and participants who were born deaf and received a CI at ages ranging from about 1 year old to early adulthood (Ear Hear. 2002;23[6]:532). Children who received their CIs during the first three years of life had P1 latencies that were similar to children with normal auditory function (Fig. 9). In contrast, children who received their CIs at 7 years of age or older had P1 latencies that were almost invariably later than their age-matched peers with normal hearing. Most (but not all) of the children who received their CIs between 4 and 7 years old also had delayed P1 latencies. Dr. Sharma concluded that the latency of the P1 component was a biomarker of auditory brain development, with later latencies representing a decoupling of the primary and secondary auditory cortices. In short, Dr. Sharma’s P1 latencies provided an electrophysiologic indication of the critical period of language development, which has long been considered to span over the first two to three years of life.

The functional implication of Dr. Sharma’s work is obvious. Children with hearing loss must be appropriately fitted with hearing technology (e.g., hearing aids or a CI) as early as possible to avoid auditory deprivation and provide access to a rich and robust model of intelligible speech. Early fitting of technology is necessary to feed the auditory cortex with...
adequate stimulation to promote synaptogenesis between the primary and secondary cortices and establish the functional neural networks between the secondary auditory cortex and the rest of the brain. We know that the typical child from an affluent home hears 46 million intelligible words by his or her fourth birthday (American Educator, 2003). These 46 million words serve as the bricks and mortar that lay the functional pathways between the primary and secondary auditory cortices and establish the neural networks necessary for sound to come to life and possess higher-order meaning. Admittedly, it is a daunting goal to provide access to these 46 million words by the fourth birthday. To do so, we must remind ourselves that every day within the critical period is an important opportunity to nourish the developing auditory brain with intelligible speech.

Figure 8. Single-unit neural responses to auditory stimulation recorded with a microelectrode inserted at different layers within the cortex of cats. Figure 8a provides an example of responses from normal hearing cats. Of note, robust neural responses were recorded at all six cortical layers. Figure 8b provides an example of responses obtained from a deaf white cat that received a CI after the critical period of auditory brain development. Note that robust responses are only observed in layers I through IV, a finding associated with decoupling of primary and secondary auditory cortex (Cereb Cortex. 2000;10[7]:714).

2. Upping the ante

Important work by Mortensen and colleagues has shown that the complex neural networks that arise when the secondary auditory cortex shares an auditory signal with other areas of the brain extends beyond the perspective of auditory skill development (Neuroimage. 2006;31[2]:842). Mortensen used PET scan to image the brain while high-performing and poor-performing CI users listened to running speech. The high performers showed activity in the left inferior prefrontal cortex while the poor performers did not (Fig. 10). The left inferior prefrontal cortex, a region often referred to as Broca's area, is involved with phonological processing, phonemic awareness, speech production, and literacy aptitude. As a result, robust connections must be developed between the primary and secondary auditory cortices so that the latter may facilitate responses in the left inferior prefrontal cortex, which is imperative for several reasons. Engaging the left inferior prefrontal cortex in response to meaningful sounds is necessary so that the child may learn to produce intelligible speech. As we've known for quite some time, children speak as they hear, and access to intelligible words is necessary to develop intelligible speech. Furthermore, access to intelligible speech is necessary to develop phonemic awareness (e.g., The “A” says “ah”), which serves as the foundation for reading development. To summarize, children with hearing loss need brain access to intelligible speech as early and as much as possible to develop their auditory skills as well as their speech production and literacy abilities.

1. The auditory brain is hungry! Feed it clearly and frequently.

So what do we do to promote auditory brain development in children with hearing loss in order to promote optimization of listening,
spoken language, and literacy abilities? We stick with the tried and true fundamentals of modern, evidence-based pediatric hearing health care. We seek to accurately diagnose children with hearing loss by 1 month of age so that hearing aids may be fitted using probe microphone measures and evidence-based targets (e.g., Desired Sensation Level 5.0/NAL-NL2) as soon as possible. For children with severe to profound hearing loss, we move forward with cochlear implantation between 6 and 9 months of age. For all children using hearing technology, we also ensure they are fitted with digital adaptive remote microphone systems (RM) so they have access to intelligible speech in our noisy world. We are convinced that the road to 46 million words is much more manageable to navigate with the use of a digital adaptive RM system. Research has clearly shown that RM technology is the most effective means to improve communication in noise (J Am Acad Audiol. 2013; 24[8]:714; Am J Audiol. 2015; 24[3]:440). Additionally, we must routinely administer audiological evaluations to ensure children are hearing well with their hearing technology.

We also must make certain that each of the child’s caregivers realizes the importance of using a hearing aid, CI, and RM technology during all waking hours from the first day when these technologies are fitted.

Finally, we must assist the family with creating a robust daily conversational/language model rich in intelligible speech. Families must also be aware that their child needs to hear 46 million words by the time he/she is 4 years old and that their auditory brain development depends upon it. Families must understand that a child’s long-term listening, spoken language, literacy, academic, and social development is influenced by early brain access to intelligible speech, and they must be equipped with skills to optimize the child’s exposure to intelligible speech. Audiologists and speech-language pathologists must work hand-in-hand with families to achieve these goals.

Neuroscientists from around the world have enlightened our profession on the neurophysiologic underpinnings of listening and spoken language development. Of particular note, children must have access to intelligible speech and meaningful acoustic input to fully develop the auditory areas of the brain and optimize spoken language and literacy aptitude. Visual stimulation in the form of sign language does not promote development of the functional synaptic connections between the primary and secondary auditory cortices. These connections serve as the springboard for a neural network/connectome that fully engages the brain and is necessary for the development of typical listening and spoken language abilities. Modern hearing technology (e.g., hearing aids, CIs, digital adaptive remote microphone systems) allows almost every child with hearing loss the access needed to fully develop the auditory areas of the brain. It is our job as pediatric hearing health care professionals to provide the children we serve with the brain growth they deserve. 

Figure 9. Cortical auditory evoked potentials (CAEP) obtained from congenitally-deafened children who received a CI prior to 3.5 years of age (red circle), 3.6 to 6.5 years of age (blue x), and of after 7 years of age (black triangles). The data points are plotted relative to the 95 confidence interval lines representing typical P1 CAEP latencies of children with normal hearing (Ear Hear. 2002;23[6]:532).

Figure 10. PET scan imaging showing typical response of (a) high-performing CI users with broad activity present in primary and secondary auditory cortex and also in left inferior frontal cortex in response to auditory stimulation from a CI, and (b) poor-performing CI users with limited activity present in primary and secondary auditory cortex and no activity in left inferior frontal cortex in response to auditory stimulation from a CI (Neuroimage. 2006;31[2]:842).