Current considerations on neural development and hearing loss in young children

BY DIMITY DORNAN

The young child's brain has the ability to change in response to new stimuli, resulting in learning, the foundation of adaptive and intelligent behaviour. For children with hearing loss, a reduction or lack of auditory stimuli can have a 'lifelong effect on many levels of brain function' particularly for those born with hearing loss [1]. However, the current convergence of advances in neuroscience, technology and the internet are transforming the potential outcomes we can provide for children with hearing loss and for their parents.

These three global influences have provided an unprecedented potential for the vast majority of very young children even those with profound hearing loss to develop clear listening and spoken language. Moreover, for parents and professionals, this convergence of advances has provided increased knowledge of the neurobiological and neurocognitive foundation for learning listening and spoken language, an upsurge in diagnostic and hearing technology, and the benefits of the internet for acquiring and sharing knowledge.

Fundamental to maximising the possible outcomes for children with hearing loss is the neurodevelopmental need to access the child's auditory brain early through newborn hearing screening, fitting of optimal hearing technology and the application of brain-centred training techniques for the development and maturation of auditory neural areas. Receiving these interventions by six months of age provides a child with maximum opportunity to take advantage of heightened neural development [2].

> **"The changing ability to rewire the brain through meaningful speech experiences in a social environment sets the foundation for later language learning."**

Auditory development in utero

Auditory brain development depends on biology, auditory experience and functional spoken language use in a social environment.

Recent evidence has spotlighted neural development in utero as the foundations of the exquisite ability to hear and discriminate sounds and relies on the precise organisation of this highly ordered structure of intricate neural connections [3]. This complex process is only just starting to be understood.

However, even before the cochlear can reliably respond to sound around 20 weeks of gestation [4], the 'quiescent and immature hair cells' in the cochlear are capable of generating spontaneous calcium-based action potentials promoting transmitter release, which drive action potential firing in developing spiral ganglion neurones [3]. This spontaneous electrical activity is thought to be significant for guiding and refining the initial phases of development of the auditory circuits. Subsequently, neuronal connections undergo a period of experience-driven refinement caused by sound-evoked electrical activity in the auditory system that eventually leads to mature, adult neural connections [5].

For babies with normal hearing in utero, a large amount of auditory brain development takes place via their muffled early hearing. The neural basis of fetal learning has been demonstrated by evidence of neural commitment specifically tuned to the speech features heard before birth and also to their memory representations. Partanen, et al [6] have demonstrated that newborns have been found to react differently to familiar sounds compared with unfamiliar sounds and found a relationship with the direct neural correlates of human fetal learning of speech-like auditory stimuli. They have found a significant correlation between higher prenatal exposure and greater brain activity. Moreover, this learning was generalised to other similar speech sounds, indicating a level of complexity of neural development before birth.

Furthermore, studies of cerebral structural connectivity in utero have recently demonstrated a 'rich-club' of interconnected hubs, a high-cost, high capacity infrastructure thought to enable efficient network communication. These are present at 30 weeks of gestation and continue to develop until birth as a proliferation of connections between core hubs and the rest of the brain [7]. This 'rich-club' organisation in the human brain precedes the later emergence of complex neurobiological function, and neural influences during this time may impact negatively on neuroplasticity and on subsequent neurodevelopment.

Once the baby is born, auditory learning involves the strengthening of these important neural long-term memory traces resulting in the improvement of discrimination skills, and particularly those forming the prerequisites for speech perception and understanding of speech [6]. Recent animal evidence for the neuroplasticity of auditory brain pathways has shown that brain cells in the auditory centres of newborn macaque monkeys showed considerably greater potential for extended growth and wiring of the auditory brain areas compared to visual cortical areas [8]. Postnatal studies of spinogenesis, dendrite growth, axon growth and electrophysiology have shown that brain cells have different growth profiles according to the area they represent, and all can vary independently of each other [9]. This data puts the historical view of neural development in context, as we used to think there was a uniformity in pyramidal cell structure. However, cells from auditory and visual areas do not start off and follow the same trajectory of growth, but have been found to have individual growth profiles [9]. Pyramidal cells play a key role in cognitive functions such as perception, reasoning, remembering, thinking and understanding.

If stimulated, the auditory brain continues to flourish, etching neural pathways permanently, with 'pre-wiring' that is not used diminished over time until it is no longer available for stimulation. The time span for maximum neural connecting and pruning, or heightened neuroplasticity, is the optimal development period for learning listening and spoken language [10].

It is highly likely that there is more than one optimal developmental period for development of listening and speaking in the central auditory system [11]. Kuhl [12] reports that the optimal development period for phonetic learning occurs prior to the end of the first year of life, whereas syntactic learning flourishes between 18 - 36 months, and vocabulary development 'explodes' at 18 months. A goal of future research is to identify the optimal development periods for phonological, lexical and grammatical levels of language.

These periods are not absolute, for example, Werker and Tees [13] have developed a nested, cascaded model of optimal periods for learning to read, with onset and offset of each period being variable, each stage building on the previous ones. Certain stages of neural development at the stage of time-sensitive readiness for learning based on perception and amenability to experience-driven change contributes to the complexity of brain structure [14].

During the first year of life, there is a window of perceptual re-organisation in the auditory area of the baby's brain [15], with a child's ability to discriminate between sounds of their own language and another language, reducing later in the first year (for monolingual babies). This refinement in listening skills is further illustrated by a decline in those aspects of speech not pivotal for processing native language [12], a gain in sensitivities pertinent to native language processing [16], and reduced ability to discriminate between sounds of their own language and another language [15].

Mapping in progress.

At birth, a baby born with significant hearing loss has already missed out on around 20 weeks of auditory stimulation and significant activation of auditory neural pathways, and is starting out to learn listening and language with a very different neurological foundation. Sound deprivation has been shown to alter the brainstem in animals and causes dendritic abnormalities in the perinatal period [17]. Neural development is affected by the timing, quality and quantity of auditory stimulation. If any of these are out of order, the brain compensates and cortical re-organisation takes place.

Untreated hearing loss causes the child's brain to be structurally and functionally different, because lack of auditory stimulation interferes with development of normal cortical grey matter and white matter maturation in primary auditory areas resulting in abnormal cortical development in the primary auditory cortex [18]. This is because delay in or lack of stimulation of auditory brain results in diminished capacity for auditory brain growth matter [19]. Poor infant speech perception can predict later language delay [20].

If the auditory brain is to develop to its full potential by capitalising on the period of optimal development, hearing loss in babies may be considered a 'neurological emergency'. Early diagnosis and early fitting of optimal hearing device followed by brain-training with a strong emphasis on abundant, meaningful, interesting speech input is critical for maximising outcomes.

The use of the latency of the P1 component of the cortical evoked response to sound varies as a function of age and can be used as a biomarker for maturation of central auditory pathways [19].

For children with severe or profound hearing loss, cochlear implants can ameliorate the deficits in the auditory system and promote cortical maturation [21]. Implantation should be as early as possible to prevent cross-modal take-over of auditory regions and rehabilitation strategies may be more effective if they target general cognitive functions instead of specialised circuits dedicated to auditory and audiovisual pattern recognition [22, 1].

The central auditory system is highly plastic for a period of about three and a half years from birth. Sharma et al [23] found that there is a sensitive period of three to four years after birth in which cochlear implantation of a child with hearing loss occurs in a highly plastic central auditory system, and that after seven years of age, the central auditory system is already re-organised and is not nearly as able to adapt to and process auditory information. This is reflected in cochlear implant outcome findings in which children implanted under ages three to four years show significantly better speech perception and language skills compared to children implanted after ages of six to seven years [24].

Finally, parent choice of an education option at birth does not result in similar outcomes for all deaf children because not all education options focus on rapid development of auditory brain pathways through a listening emphasis.

The question is, are the changes in neural architecture caused by hearing loss reversible?

This is a topic of ongoing investigation. We know that scientists have the ability to restore abnormal synaptic structure in auditory nerve endings of deaf cats by stimulating auditory nerves for three months with six channel cochlear implants [21]. Moreover, it has been shown that auditory stimulation can facilitate preservation of auditory brain structures and reverse the effects of auditory deprivation [25]. We also know that cochlear implant <12 months is optimal [26], and implantation after seven years occurs into a reorganised central auditory system [25]. However we do know that children with significant hearing loss are able to progress at the same developmental rate for listening, speech and language development and catch up with normally hearing peers [27].

This is most likely if children are implanted early (<12 months), use oral communication [26], and parents are trained to provide an enriched auditory environment, one in which the frequency, quality and quantity of meaningful auditory input targets the development of auditory brain pathways [27].

References

1. Kral A, O'Donoghue GM. Profound deafness in childhood. *N Engl J Med* 2010;**363**:1438-50.
2. Joint Committee on Infant Hearing Position Statement (JCIH) 2007. Accessed 5 Oct 2014 at http://pediatrics.aappublications.org/content/120/4/898.full.pdf+html
3. Kennedy HJ. New developments in understanding the mechanisms and function of spontaneous electrical activity in the developing mammalian auditory system. *J Assoc Res Otolaryn* 2012;**13**:437-45.
4. Rubel EW. Strategies and problems for future studies of auditory development. *Acta Otolaryngol* (Stockh.) 1985;**421**:114-28.
5. Blankenship AG, Feller MB. Mechanisms underlying spontaneous patterned activity in developing neural circuits. *Nat Rev Neurosci* 2010;**11(1)**:18-29.
6. Partanen E, Kujala T, Näätänen R, et al. Learning-induced neural plasticity of speech processing before birth. *Proc Nat Acad of Sci* 2013;110:15145-50.
7. Ball G, et al. Rich-club organization of the newborn human brain. *Proc Nat Acad of Sci* 2014;**111**:7456-61.
8. Elston GN, Okamot T, Oga T, Dornan D, Fujita I. Spinogenesis and pruning in the primary auditory cortex of the macaque monkey (Macaca fascicularis): an intracellular injection study of layer III pyramidal cells. *Brain Res* 2010b;**1316**:35-42.
9. Elston GN, Fujita I. Pyramidal cell development: postnatal spinogenesis, dendritic growth, axon growth, and electrophysiology. *Front Neuroanat* 2014;**8**:78.
10. Newport EL, Bavelier D, Neville HJ. *Language, Brain, and Cognitive Development: essays in honor of Jacques Mehler*, Cambridge, MA, US: The MIT Press 2001.
11. Lee DS, Lee JS, Oh SH et al. Cross-modal plasticity and cochlear implants. *Nature* 2001;**409(6817)**:149-50.
12. Kuhl PK, Williams KA, Lacerda F et al. Linguistic experience alters phonetic perception in infants by 6 months of age. *Science* 1992;**255(5044)**:606-8.
13. Werker J. Speech perception as a window for understanding plasticity and commitment in language systems of the brain. *Developmental Psychobiology* 2005;(0012-1630);**46(3)**:233.
14. Markman, et al. Language development after cochlear implantation: an epigenetic model. *J Neurodev Disord* 2011;**3(4)**:388-404.
15. Werker JF, Tees RC. Cross-Language speech perception: Evidence for perceptual reoganisation during the first year of life. *Infant BehDevel* 1984;**7**:49-63.
16. Jusczyk PW. Some beginnings of word comprehension in 6 month-olds. *Psychol Sci* 1999;**10**:172-5.
17. Moore JK, Linthicum FH Jr. The human auditory system: A timeline of development. *Int J Audiol* 2007;**46**:460-78.
18. Smith, et al. Morphometric Differences in the Heschl's Gyrus of Hearing Impaired and Normal Hearing Infants. *Cereb Cortex* 2011;**21(5)**:991-8.
19. Sharma A, Dorman MF, Kral A. The influence of a sensitive period on central auditory development in children with unilateral and bilateral cochlear implants. *Hearing Res* 2005;**203(1-2)**:134-43.
20. Molfese DL. Predicting dyslexia at 8 years of age using neonatal brain responses. *Brain Lang* 2000;**72**:238-45.
21. Ryugo DK, Kretzmer EA, Niparko JK. Restoration of auditory nerve synapses in cats by cochlear implants. *Science* 2005;**310**:1490-2.
22. Lee H-J, et al. Cortical activity at rest predicts cochlear implantation outcome. *Cereb Cortex* 2007;**17**:909-17.
23. Sharma A, et al. Central auditory maturation and babbling development in infants with cochlear implants. *Arch Otolaryngol Head Neck Surg* 2004;**130(5)**:511-16.
24. Gantz BJ, Rubinstein JT, Tyler RS, et al. Long-term results of cochlear implants in children with residual hearing. *Ann Otol, Rhinol Laryn* 2000;**109**:33-6.
25. Matsushima J, Shepherd RK, Lee Seldon H, et al. Electrical stimulation of the auditory nerve in Kittens: Effects on cochlear nucleus morphology. *Hearing Res* 1991;**56(1-2)**:133-42.
26. Leigh J, Dettman S, Dowell R, Briggs R. Communication development in children who receive cochlear implant by 12 months of age. *Otology Neuro Otology* 2013;**34**,3:443-50.
27. Dornan D, Hickson L, Murdoch B, et al. Is Auditory-Verbal Therapy effective for children with hearing loss? *The Volta Review* 2010;**110(3)**:361-84.

Dimity Dornan,

Associate Professor, Executive Director and Founder Hear and Say, Auchenflower, Toowong QLD, Australia

E: dimity@hearandsay.com.au
W: www.hearandsay.com.au

Declaration of competing interests
None declared