Factors behind the 'cleaning' of the auditory pathway in late implantation of prelingual oral deaf adults

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Pre- and peri-lingually deaf adults are benefiting from late cochlear implantation. While much has been written about the emotional experiences, we review auditory plasticity based on 16 months of CI usage by the first author. We suggest that the goal of speech discrimination in quiet via bimodal hearing may accrue from some or all of the following: 1) amplification of low-frequency sounds since infancy, 2) usage of residual hearing via parent-child interaction in auditory training, 3) improved synaptic contact via spike activity from high stimulation rates fused with natural firing from residual hair cells, 4) exposure to singing and music as infants, 5) top-down linguistic processing, 6) reduced cognitive load, 7) episodes of sleep-induced tinnitus-like symptoms after a period of intense auditory exposure, 8) auditory exposure throughout the day, 9) based on inference of imaging scans of 5 oral deaf adults, the distribution of the gray matter cortical thickness of the Heschl's Gyrus (HG) as well as the spatial topography of the acoustic radiation white matter tract from the thalamus to the HG appear to be maintained, and 10) auditory training for bottom-up phoneme processing and auditory working memory.

INTRODUCTION

An increasing number of pre- and peri-lingual oral deaf people get cochlear implants late as adults. This may account for adults age 30-49 years old being the second largest group receiving CI nationally in the United Kingdom (see Fig. 4 in Raine, 2013) and locally at Johns Hopkins Hospital in the past five years. A contributing factor is the decreasing benefit of hearing aids (HAs) due to aging. Evidence suggests that prior use of HAs can provide positive outcomes for pre/peri-lingual deaf late implanted adults (PLDLI) (Caposecco *et al.*, 2012). The purpose of this review is to discuss factors contributing towards the goal of speech discrimination in quiet via bimodal hearing from the perspective of a PLDLI auditory scientist.

LOW-FREQUENCY RESIDUAL HEARING SINCE INFANCY

It has long been observed that successfully mainstreamed deaf adults had usable low-frequency residual hearing (Urbantschitsch and Goldstein, 1898; Bárczi, 1936;

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Ewing and Ewing, 1938). Then the advent of the transistor resulted in a remarkable example of serendipity with different people making similar observations in the same period (Hardy *et al.*, 1951; Huizing and Pollack, 1951; Wedenberg, 1951; Beebe, 1953; Whetnall, 1956; Guberina, 1957): Deaf children were able to communicate clearly and naturally. These observations were synthesized in the 1970s via the 'Ling Six Sounds' as a means to test the potential ability of the deaf child to comprehend sounds (Ling, 2002). The Ling Six Sounds consist of m, oo, ee, ah, sh, and s, which are essential to speech and language development. The first four are in the low-to-mid frequency range and the last two in the moderate-to-high frequency range. Early diagnosis of hearing loss followed by intervention with HAs probably accounts for the maturation of P1 latency in cortical auditory evoked potentials (CAEP) of babies fitted with HA (Nash *et al.*, 2007). This suggests that HA amplification of low-frequency sounds within 20 dB of normal hearing allows a critical part of the 'speech banana' to be perceived by the deafened brain. Also the proximity of the parent in communicating with the deaf child is critical in facilitating 'motherese' (Brown *et al.*, 2001) especially within the first three years critical for the maturation of the P1 latency (Sharma *et al.*, 2002). This leads to two questions. First, how can low-frequency input with HA enable suprasegmental discrimination in speech production and understanding (Abberton and Fourcin, 1978) and second, why do PLDLI born before the digital HA era take longer to reach 'reasonable' speech discrimination levels with CI.

EXPOSURE TO SINGING AND MUSIC AS INFANTS

The spectrum of music is much larger than that of speech, so it is possible that lullabies and nursery rhymes generate low-frequency tones. In turn this may have a positive effect on suprasegmental development seen in CI subjects who began with HA (Most and Peled, 2007). This reinforces the suggestion that access to acoustic hearing, especially low-frequency tones, creates a foundation for music perception accessed later with CI (Hopyan *et al.*, 2012). That music sounds more pleasant (Fuller *et al.*, 2013) may be attributed to the electrical stimulation of the auditory pathways previously developed by acoustic stimulation (Hopyan *et al.*, 2012). There is evidence (Fawkes and Ratnanather, 2009) to show that at one extreme singing improves cadence in deaf children and at the other extreme deaf children are capable of studying and performing music as an academic subject to a very high standard. This leads to the question whether it would be helpful to prime the auditory pathway with HAs via speech and music for a few months prior to CI.

FUSING SYNAPTIC CONTACT AND SPIKE ACTIVITY

HAs can do only so much, i.e., low-frequency amplification to within 20 dB of normal hearing. But the high stimulation rate via CI results in increased synaptic contact and activity at every stage of the auditory pathway from the spiral ganglion cells to the auditory cortex (Kral *et al.*, 2000; Chen *et al.*, 2010). For example, the density of synaptic contacts correlates with the spike activity (O'Neil *et al.*, 2011). So bimodal hearing fuses the natural spike activity from the HA at low frequency with that from the CI at all frequencies to give ‘warmth’ and ‘clarity’, respectively (Crew *et al.*, 2013). It could be argued that the high stimulation rate from the CI induces the natural ‘stochasticity’ properties of the auditory pathway but there is no correlation of stimulation rate with speech perception (Shannon *et al.*, 2011). It is interesting that the parietal cortex activity is significant in deaf adults who have not used HAs (Gilley *et al.*, 2008). This may relate to the initial moments of activation in which the CI patient experiences whole body sensation via the homunculus distribution along the motor cortex. This suggests that the distribution of receptors in the thalamocortical network might be affected by rate-level functions (Metherate *et al.*, 1990). Hence the question whether white-matter tracts to the auditory cortex from the thalamus tolerate high spike activity while tracts to the parietal cortex cannot.

SLEEP-INDUCED PLASTICITY

There have been anecdotal reports that following intense periods of auditory exposure soon after CI activation, ‘whooshing’ brain waves have been experienced either before or at the end of deep REM sleep or both. It is unlikely to be tinnitus as it did not cause distress nor did it appear to be vascular. It is episodic hence being mentioned online by CI subjects. These events could be signs of ‘sleep-induced’ plasticity. It is known that sleep consolidates experience-dependent plasticity (Aton *et al.*, 2013), so do these waves reflect cortical protein synthesis, remodelling of neurons and synapses (Kral, 2013) or the fast propagating waves observed by Reimer *et al.* (2011)? That these episodes do not occur after some time suggests that adaptation has taken place, i.e., it is a ‘positive’ signature of the adapting brain.

TOP-DOWN PROCESSING

PLDLI have already acquired good language so it is not surprising that top-down linguistic processing via contextual analysis is maximised. This top-down cognitive processing mechanism is probably an adaptation of that used in speechreading (Capek *et al.*, 2008). Also the brain is Bayesian, i.e., utilises probabilistic inference (Shannon *et al.*, 1995; Boothroyd, 2010) which suggests that adaptive learning algorithms could be implemented in CI processors. Despite excellent open-set speech recognition scores, the long term challenge is auditory working memory which leads to the question whether this is similar to understanding speech in noise.

REDUCED COGNITIVE LOAD

There is an increase in multi-tasking such as listening to audiobook, podcast, or radio while working on a computer or watching TV; similarly passive listening in meetings is possible. Anecdotal reports of deaf adults who did not use HAs and used signing prior to CI and found it difficult to get ‘over the hump’ in open-set speech suggest that cross-modal plasticity prior to CI may be difficult to unravel. This may be explained by functional MRI studies of deaf adults which showed that those who do not sign process information differently from those who do (Cardin *et al.*, 2013). A key network is the connection between the frontal cortex including Broca’s responsible for executive function and the temporal cortex including association cortex such as the planum temporale (PT) responsible for speech and language processing. The extensive cross-modal plasticity via the takeover of the auditory cortex by visual processing suggests that the acoustic radiation (AR) from the medial geniculate body in the thalamus to the auditory cortex and the optic radiation (OR) from the lateral geniculate body in the thalamus to the visual cortex may cross, overlap, or fuse in deaf adults. It is also challenging to visualise the AR and OR in whole-brain diffusion tensor images (DTI) but Fig. 1 shows it is possible to generate AR via probabilistic based white-matter (WM) fibertracking (Ratnanather *et al.*, 2013) and that spatial topography of the AR is similar in deaf and normal subjects. It is also possible to show that WM tracts from the Heschl's Gyrus (HG) to the posterior region of the PT passes through the OR. This leads to the question whether the CI can "uncouple" the OR and AR.

AUDITORY CORTEX

The thickness of the cortical mantle differs in motor and sensory cortices (Jones, 2004). Figure 2 shows histograms of distances of gray matter (GM) relative to four different cortical surfaces related to hearing, speech, and language from MRI scans from groups of PLDLI adults (prior to CI) and controls. Following Kral and Eggermont (2007), these histograms can be interpreted with respect to upper and deep cortical layers. The differences further from the GM/WM surface could be related to the lack of lateral input that affects bottom-up processing seen in deaf subjects; the similarities closer to the surface could be related to top-down processing in deaf adults with normal CAEP latencies. This leads to the question whether HAs help to prime the auditory pathway and synaptogenesis in the upper layers can be facilitated by auditory training.

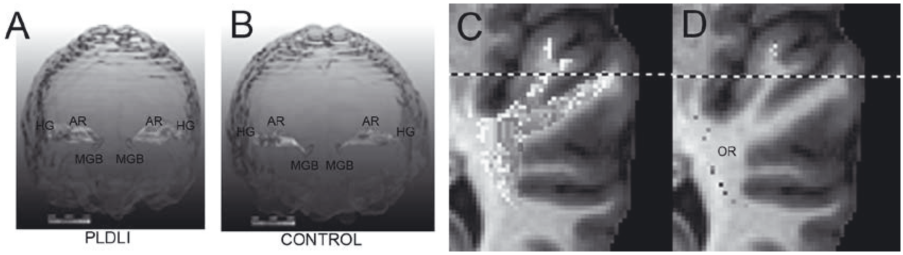

Fig. 1: Acoustic radiation WM tracts from the MGB to the HG in one deaf (A) and one control (B) subject. WM tracts from the HG to the PT (C) and those that pass through the optic radiation (D) terminate in the posterior PT.

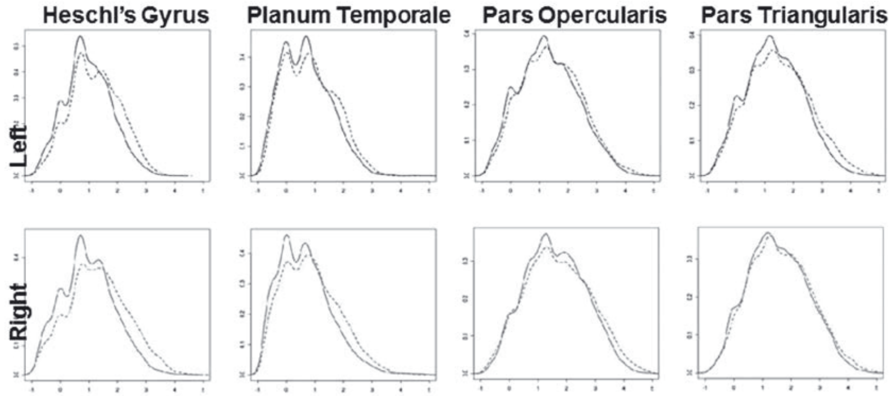

Fig. 2: Normalised distances of GM from cortical surfaces related to hearing, speech, and language. Solid and dashed curves respectively correspond to pooled groups of five age- and gender-matched normal and deaf adults. KS-tests show thinning in deaf group ($p < 0.0001$).

AUDITORY EXPOSURE

Anecdotal reports of low CI usage are difficult to believe. What is the point of the CI if it is not consistently used? (Gordon *et al.*, 2011) First it is suggested that adaptation to noise would be quicker if digital HAs were used prior to CI. It is likely that in the deafened brain, it takes longer to mask noise. So it is important not to let background noise dominate the CI during waking hours. However auditory training in noise is said to improve neural timing (Song *et al.*, 2012). So in the early days, it is helpful to self-test with the Ling Six Sounds iPad app as well as the Virtual Piano software. In comparison with autobiographical accounts which have generally been emotional, perhaps the most substantive account is that by someone forty years after benefiting from auditory training as a child (Beebe, 1953; Younglof, 1997). The HA in the contralateral ear should be used after about eight weeks so that the benefits from bimodal hearing can be accrued. This leads to the question how loudness balance between CI and HA can be optimised (Neuman and Svirsky, 2013).

AUDITORY TRAINING

Auditory training (AT) should begin with bottom-up processing, i.e., acoustic analysis of speech starting with suprasegmental sounds – vowel (V), consonant (C), and consonant-vowel (CV) – and then progressing to CVC and CVCVC. In top-down processing, cognitive analysis is used to extract meaning so it is important not to ‘think’ but to learn to process phonemes. Among the existing AT software, Angel Sound (Fu and Galvin, 2012) is freely available. However there is variation in provision of AT in clinics in the US (Sorkin, 2013) and the UK (Raine, 2013). One solution is to develop adaptive learning tablet applications with feedback which might allow for monthly visits by more patients instead of few weekly visits. In future, music should be incorporated as part of AT as it can improve neural timing (Kraus and Chandrasekaran, 2010).

SUMMARY

For PLDLI adults, it is a work in progress but with new experiences accrued almost on a daily basis. According to Michael Dorman, who was involved in the early days of multichannel CI research, if there is a way to 100% speech recognition in quiet then there are several ways of getting there. Granted that technological developments in CI have begun to mature, it is necessary to develop effective strategies for AT to maximise benefit from the remarkable ability of the brain to adapt to the new auditory input. However, is it reasonable to infer what might happen in PLDLI from anatomical changes in completely deafened animals? MRI, DTI, and functional MRI (fMRI) scans prior to CI could be useful tools for customizing surgical and rehabilitative strategies, but post-operative PET imaging presents challenges. Last but not least, fMRI studies of brain activity via HA amplification are possible without electromagnetic interference from HA (Ratnanather et al., in preparation) and may provide clues for substrates of PLDLI.

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