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Cortical auditory evoked responses from an implanted ear after 50 years of profound unilateral deafness

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ABSTRACT A male with unilateral deafness in the right ear since 8 years of age developed a sudden hearing loss in the left ear at age 63. A hearing aid was fitted in the left ear with limited benefit. The right ear received a cochlear implant (CI) 20 months later. Cortical auditory evoked potentials (CAEPs) and speech recognition scores (SRS) were measured in free-field three, six and nine months after implantation with the hearing aid alone, CI alone and bimodal condition (hearing aid and CI together). Three months after implantation the cortical auditory deprivation. CAEPs measured over time show evidence of binaural interaction and improvements in SRS. Copyright © 2007 John Wiley & Sons, Ltd.

Keywords: unilateral deafness; auditory deprivation; cortical plasticity; bimodal condition; binaural interaction

Introduction

When the auditory system is not stimulated with sound for a long period of time, reorganisation of the frequency maps in the auditory cortex occurs, as well as alterations in neural responses and binaural interactions at various levels in the auditory pathways (e.g. Ponton and Eggermont, 2001; Eggermont et al., 1997). Recently, cortical auditory evoked potentials (CAEPs) have been used to investigate auditory plasticity in patients with long-duration deafness that then have some hearing function restored using a cochlear implant (CI). It has been demonstrated that in such individuals there is enhancement of cortical response amplitude and morphology over time, which corresponds well with improvements in behavioural speech recognition scores (SRS) (Purdy et al., 2001). These improvements are thought to reflect plasticity of auditory function with the reintroduction of auditory input over time (Ponton and Eggermont, 2001).

Unilateral hearing loss is also thought to alter the neuronal activation and binaural interactions in the auditory pathways, however; this has been studied in less detail. Khosla et al. (2003) found that reduction in ipsilateral-contralateral amplitude differences for N1-P2 CAEP occurred in patients with profound left ear unilateral deafness. This finding suggests there is reorganisation in the auditory cortex in unilateral left deafness, with cortical activation increasing in the left hemisphere. In contrast, patients with unilateral right deafness did not show evidence of reduced ipsilateral-contralateral amplitude differences. This suggests there is less compensatory plasticity increase in activation of the left hemisphere with deafness in the right ear alone.

It is of interest to investigate the effect of reintroducing auditory input in individuals that are implanted after a long period of profound unilateral deafness to determine whether some or all aspects of normal cortical function are restored.

This paper presents a case study of a 64 year old male (M.M.) with a profound sensorineural hearing loss in the right ear since childhood and a later onset fluctuating moderate-severe sensorineural hearing loss in the left ear. The duration of profound hearing loss in the right ear was longer than 50 years and was attributed to mumps in early childhood. M.M. first became aware of his profound hearing loss in the right ear at school when he was 8 years old. In 2001 M.M. had a sudden hearing loss in the left ear, which was diagnosed as secondary endolymphatic hydrops. Computerized Tomography scans of the temporal bones revealed an asymmetry in size of the cochlear aqueducts, the left being larger than the right.

M.M. was referred to the audiologist for hearing aid assessment after the episode of sudden left hearing loss. A hearing aid (Phonak Claro 211 daz) was fitted in the left ear. Hearing levels in the left ear however continued to fluctuate, making it difficult to programme the hearing aid. The ear mould was also an ongoing problem as it was usually uncomfortable due to the tight fit needed to reduce acoustic feedback.

M.M. was not considered a suitable candidate for a CI as audiological assessment showed aided SRS of 95% with his left hearing aid in free-field using Central

Institute for the Deaf sentences. This result is above CI candidacy guidelines, which recommend aided SRS in quiet worse than 70% as a criterion for implantation (Dowell et al., 2003). Furthermore, the right ear had not had auditory stimulation for over 50 years, which is traditionally a contra-indication for implantation. In spite of this the surgeon and M.M. agreed that an implant in the right ear would be attempted, as there was 'nothing to lose'. Consequently the right ear was implanted in August 2002 with a Cochlear Nucleus-24 contour. In September 2002 the implant was mapped and switched on using an Esprit 3G speech processor implementing Advanced Combination Encoder strategy.

This study shows the changes in M.M.'s auditory responses with the CI in the right ear and hearing aid in the left ear during the first nine months after implantation. Cortical auditory evoked potentials were recorded using a high frequency stimulus before and after implantation. The stimulus was selected based on previous evidence for robust cortical responses to 4 kHz tonebursts in adult CI users (Kelly et al., 2005). Changes were also expected for cortical responses in this frequency region based on previous evidence for high frequency cortical reorganisation in humans with acquired hearing loss (e.g. Dietrich et al., 2001; Thai-Van et al., 2003).

Method

Pure tone audiograms from 250 Hz to 8 kHz and SRS using Arthur Boothroyd word lists presented at maximum comfortable levels were measured under headphones several times before and after implantation.

CID sentences were presented at 65 dB SPL from a loudspeaker placed 1 m in front of the subject with no competing noise in a sound treated room to measure aided and unaided SRS pre-implantation.

City University of New York sentences were used with the same settings postimplantation to measure SRS in free-field with the CI alone, hearing aid alone and bimodal (CI and hearing aid) conditions.

CAEPs were also measured with the CI alone, hearing aid alone and bimodal (CI and hearing aid) conditions using 4 kHz tonebursts at the most comfortable loudness level via a loudspeaker at 45° azimuth and 1 m distance. The subject was seated comfortably in a recliner chair watching a self-chosen movie with subtitles and no sound to keep alert during testing.

SRS and CAEP measurements were made three, six and nine months postimplantation to show changes of the auditory pathways with reintroduction of sounds.

CAEP were carried out with a Neuroscan STIM and SCAN (version 4.2) evoked potential system, which generated the stimuli and recorded the evoked responses.

The tone burst stimuli were windowed using 20 ms linear rise and fall times, with 60 ms total duration. The inter-stimulus interval (ISI) was 1125 ms and stimuli were presented with alternating onset polarity. Stimuli were presented in

two blocks of 100 per stimulus. The choice of stimulus duration was based on previous evidence that tonal stimuli with rise times less than 30 ms and plateau times longer than 10 ms are recommended for CAEP recordings (Onishi and Davis, 1968). Robust CAEP recordings have been obtained previously in adult CI users using 20–20–20ms tone bursts (Kelly et al., 2005). The electrical artefact picked up by the recording electrodes when acoustic stimuli are delivered to the CI is related to stimulus duration and hence a relatively short stimulus duration was selected to reduce the chance of masking the components of interest in the CAEP waveform.

CAEPs have a long refractory time of several seconds (Picton et al., 1990). An ISI in the order of 1 to 2 is regarded as optimal for CAEP recording (Picton et al., 1977) while very long ISIs prolong recording times. Latency increases and amplitude reductions occur when ISI reduces to less than a second (Tremblay et al., 2005). Hence, an ISI of 1125 s was selected as a compromise to ensure robust CAEP recordings and acceptable recording times. Alternating stimulus polarity was used to reduce electromagnetic stimulus artefact pick up by the recording electrodes.

Gold cup electrodes were placed at Cz (vertex) and A2/A1 (contralateral earlobe, reference), with a ground electrode on the forehead. Electrode impedances were maintained below 5 kOhms. The vertex to contralateral ear electrode montage minimises CI electrical artefact and optimises CAEP amplitude (Sharma et al., 2002). The electrical artefact that often occurs when recording evoked responses in CI users can be five to ten times larger than the averaged cortical response (Gilley et al., 2006). Key features of the artefact that help separate it from a cortical response are that the artefact starts at zero ms and has a square onset (Gilley et al., 2006). In the current case study there was little evidence of CI artefact in the CAEP recordings.

Electroencephalography files with a recording window of -100 to +600 ms were used, including a pre-stimulus time period of 100 ms. The EEG signals were band pass filtered online (0.05–200 Hz) and low pass filtered offline (0.1–30 Hz, 24 dB/ octave slope). Artefact rejection was used to exclude responses exceeding +/- 100 μ V. Grand average CAEP waveforms were created from the two blocks for each tone.

CAEP peak amplitudes and latencies were identified by two independent observers. The amplitude of P1 was defined as the largest positive deflection occurring between 50 and 100 ms after stimulus onset. The amplitude of N1 was identified as the largest negative deflection between 80 and 120 ms after stimulus onset. P2 amplitude was defined as the largest peak occurring between 150 and 200 ms (Stapells, 2002). The latency of the peak was measured at the centre of the peak. When the waveform contained a double peak of equal amplitude or a peak with a plateau, the latency was measured at the midpoint of the peak.

Table 1: Air conduction pure tone thresholds for the left ear (NR = no response)								
	250 Hz	500 Hz	1 kHz	2 kHz	3 kHz	4 kHz	6 kHz	8 kHz
Feb 01	80	95	90	90	90	85	NR	NR
May 01	65	70	65	80	70	70	80	75
Aug 02	60	65	75	70	65	75	80	70
Mar 03	75	75	75	75	75	85	110	100
Jul 03	65	70	90	95	85	95	95	100
Aug 03	65	70	85	95	80	85	110	100
Dec 03	65	75	90	90	90	85	95	NR

Table 2: SRS results pre-operatively and three, six and nine months after implantation						
	AB words left at MCL via headphones	Hearing aid left CID or City University of New York sentences	CI right City University of New York sentences	Hearing aid plus CI City University of New York sentences		
Pre-CI 3 months post 6 months post 9 months post	60% Not tested 34% 40%	95% (CID) Not tested 7% (CUNY) 9% (CUNY)	Not tested 56% 66% 58%	Not tested 60% 47% 87%		

Results

As demonstrated in Table 1, pure tone audiometric thresholds fluctuated in the left ear consistent with the diagnosis of secondary endolymphatic hydrops, while the right ear has consistently absent responses up to 110 dB HL at all tested frequencies. The maximum fluctuation was 20 dB at a given frequency with a 5 dB variation in the four frequencies pure tone average.

As shown in Table 2, SRS were very poor in the left ear after implantation compared to pre-operative results. The hearing loss was fluctuating at this time, however, and more difficult speech material was used for post-implant testing. By nine months post-implantation the CUNY speech scores indicate that bimodal listening is superior to the CI alone.

Figure 1 provides an example of cortical responses recorded in an adult with normal hearing, showing P1, N1 and P2 at about 50, 90 and 180 ms, respectively. The difference in the bimodal condition compared to CI or hearing aid alone that was seen in the SRS results is evident in M.M.'s cortical responses at six months (Figure 2) and nine months (Figure 3) post-CI. The waveforms in Figure 3 recorded at nine months post-CI show P1 and N1 peaks similar to the results for a normal listener shown in Figure 1. P2 is smaller, however, and M.M.s' P1 and N1 latencies are slightly later than normal. They are consistent with those reported by Kelly and her colleagues (Kelly, 2001; Kelly et al., 2005), however, for adult CI users.



Figure 1: CAEP recorded to 4 kHz tonebursts (60 ms duration, 1125 ms ISI) in an adult with normal hearing. Stimuli were presented to the right or left ear via insert earphones.



Figure 2: CAEP six months post-CI. CAEP amplitude is larger in the hearing aid alone condition. Thick line: hearing aid alone; dashed line: CI alone; thin line: bimodal.

Kelly (2001) reported P1, N1 and P2 latencies for 4 kHz toneburst stimuli of 57 ms (SD 14 ms), 99 ms (SD 15 ms) and 178 ms (SD 26 ms) in adult CI users with an average of 2.9 years of CI experience. M.M.'s P1 and N1 latencies are similar to these values.



Figure 3: CAEP at nine months post-CI. CAEP amplitudes are similar in all conditions. Thick line: hearing aid alone; dashed line: CI alone; thin line: bimodal.

Comparison of the waveforms in Figures 2 and 3 indicates a change in binaural interaction, with bimodal cortical responses smaller than with hearing aid alone and slightly larger than with CI alone, at six months. At nine months the pattern of results has significantly changed. The CI alone response is now similar to the hearing aid alone condition. The amplitudes of the responses were very similar for the three different conditions but the latency of the bimodal cortical responses was later than for CI alone. The peak–to-peak amplitudes are similar between six and nine months for the hearing aid alone condition (note change in scale between Figures 2 and 3). Figure 4 shows the results of Cz recording for 4 kHz in the bimodal conditions three, six and nine months post-implantation. Figures 5 and 6 show the 4 kHz results for the CI and hearing aid alone, respectively, at three, six and nine months post-implant.

Figures 4 and 5 show that bimodal and CI cortical responses improved between three and six months and changed very little between six and nine months. Hearing aid alone responses, on the other hand, reduced over time. These results are consistent with SRS and M.M.'s subjective report of relying more and more on the CI with less perceived benefit from the hearing aid.

Conclusion

This case study shows that auditory responses can be elicited via a CI in an adult even after more than 50 years of unilateral auditory deprivation. Changes in SRS



Figure 4: CAEP bimodal (hearing aid left ear, CI right ear). The cortical responses in the bimodal condition changed over time after implantation. Thick line: three months post-CI; dashed line: six months post-CI; thin line: nine months post-CI.

over time and differences in performance between the left hearing aid, the right implant and bimodal stimulation were reflected in the CAEP results.

SRS were very poor in the left ear after implantation, but the hearing loss was fluctuating at this time and more difficult speech material was used for post-CI testing. Despite this, at nine months post-CI the CUNY speech scores indicate that bimodal listening is superior to the CI alone.

Eighteen months after implantation the subject reports great satisfaction with the implant. He uses bimodal stimulation (CI in the right ear and hearing aid in the left ear) but reports that he relies mostly on the CI. He is now back to work and reports significant improvement in hearing ability. Left hearing continued to fluctuate, typical of endolymphatic hydrops.

The difference in bimodal listening compared to CI or hearing aid alone is evident in the cortical responses at six and nine months post-CI (Figure 4). These show a binaural interaction effect, with different cortical responses in the bimodal condition than with either device alone.

This case study illustrates how cortical reorganisation is possible in a mature brain even more than 50 years after unilateral auditory deprivation. Four years after



Figure 5: CAEP CI alone. The amplitude of the cortical responses with CI alone increases over time after implantation. Thick line: three months post-CI; dashed line: six months post-CI; thin line: nine months post-CI.

implantation of the right ear M.M. showed SRS of 92% with the implant alone and 0% with the hearing aid alone. The left ear has recently been implanted and we will endeavour to report his progress in the future.

In 1991 Pelizzone and colleagues compared the post-implant cortical responses of one subject with bilateral long-term deafness, which also was longer than 50 years. The right ear was congenitally deaf while the left ear became deaf at age 7. After bilateral cochlear implantation, electrical stimulation elicited normal P1, N1 and P2 responses in the left ear with acquired deafness but abnormal responses in the congenitally deaf ear. These findings are consistent with the current study, which also showed normal P1 and N1 cortical responses after implantation in an ear with a long-term acquired deafness. Pelizzone et al.'s case had a more robust P2 response and earlier latencies than M.M. This difference may be due to the shorter duration of implant experience of M.M. (three to nine months) compared to Pelizzone's case (two years).

We also compared our subject's CAEP results to the results of an adult with normal hearing (Figure 1). The responses from M.M. at nine months after implantation were similar to those of normal hearing subject, although somewhat later



Figure 6: CAEP hearing aid alone. The amplitude of the cortical response with the hearing aid alone decreases over time after implantation. Thick line: three months post-CI; dashed line: six months post-CI; thin line: nine months post-CI.

and with reduced P2 amplitudes. This suggests that, although the right ear was deprived of auditory stimulation for more than 50 years, this ear had a well-established auditory pathway in the first 8 years of life. This is consistent with evidence that cortical responses are largely mature by about 12 years, with substantial maturational change during the preschool years (Ponton and Eggermont, 1991; Sharma et al., 2002). Deaf children receiving a CI can develop mature cortical responses, but not if implantation is in late childhood (Ponton et al., 1999).

These results have implications when predicting outcomes of cochlear implantation in adult subjects with long-term deafness. Our results confirm Pelizzone et al.'s findings, suggesting that the age of the acquired hearing loss may be more important than the time lapsed between the loss of hearing and cochlear implantation. Furthermore, pre-implant electrically evoked CAEP may be a useful test to predict outcome of CI in long-term deafness.

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