



## Cortical Deafness- A Rare Case Report

### KEYWORDS

Cortical deafness, auditory agnosia, bilateral cortical lesions, auditory evoked potentials, Speech language assessment.

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### ABSTRACT

Central Auditory deficits can vary from pure word deafness to unresponsiveness to all environmental sounds. Cortical deafness is a term used to define severe central auditory deficits following bilateral lesions which are rare and provide unique opportunity to evaluate audiological. Confirmation of cortical deafness requires detailed test battery approach with Auditory Evoked Potentials (AEP). This case report highlights the importance of AEP's along with routine audiological testing in diagnosis of cortical deafness. Also we emphasis on complete speech , language evaluation to rule out aphasia, a language disorder commonly accompanied with mild to severe auditory comprehension deficits.

### Introduction

Cortical deafness is the severe form of central auditory deficit, which refers to unresponsiveness to all sounds due to lesion in bilateral cerebral hemispheres [5]. Central deafness is exceedingly rare because each ear projects to both sides of the brain stem, thalamus, and cortex. A number of case reports on central deafness state that when the patient suddenly lost the ability to comprehend speech, he complained that he could not hear and was unresponsive to environmental sounds, music, and speech [3& 4].

### Case Report

A 24 year old, right handed male with the sudden onset of deafness for the past three weeks reported to our Department with the complaint of inability to hear any sounds and difficulty in understanding speech. On examination the patient was not responding to verbal questions or environmental sounds and did not exhibit startle to loud noises. He was alert with fluent speech and occasional literal paraphasias. He had history of an old right middle cerebral artery infarct 2 years back and recently has developed left middle cerebral artery infarct with right hemiparesis since 3 weeks. The CT scan done on 15.7.2014 showed an old right frontoparietotemporal infarct and a new left parieto-temporal infarct.

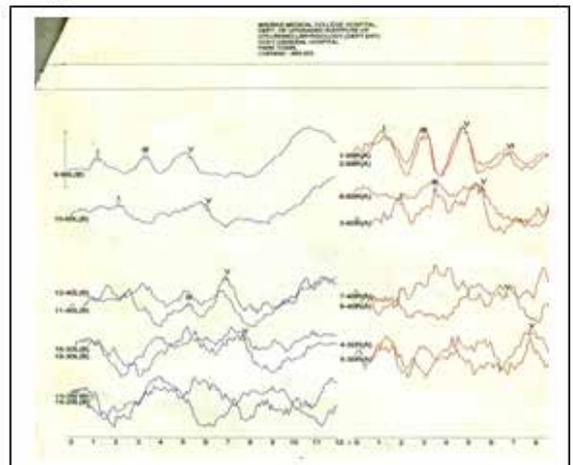
Detailed Hearing and Speech, Language Assessment was carried out at the Department of Audiology and Speech language Pathology on 1.8.2014. Hearing assessment included assessment of pure tone Audiometry, Speech audiometry, Immitance audiometry and Auditory evoked potentials. Pure tone thresholds showed bilateral moderate sensory neural hearing loss (SNHL) but the responses were inconsistent. Speech discrimination scores were poor (20%) bilaterally. 'A' type tympanogram with normal ipsilateral and contralateral stapedial reflexes were present in immitance audiometry. Auditory brainstem evoked potentials recorded with click stimuli at repetition rate of 30.1 per second showed wave I, III and V peaks within normal latency and amplitude bilaterally till 30dBnHL intensity level. Cortical evoked potentials for Short duration speech stimuli /ba/ showed absence of late latency responses (LLR) in

right ear and left ear. Middle latency responses (MLR) with Pa and Na peaks below 50ms latency were present in left ear alone (Figure 1).

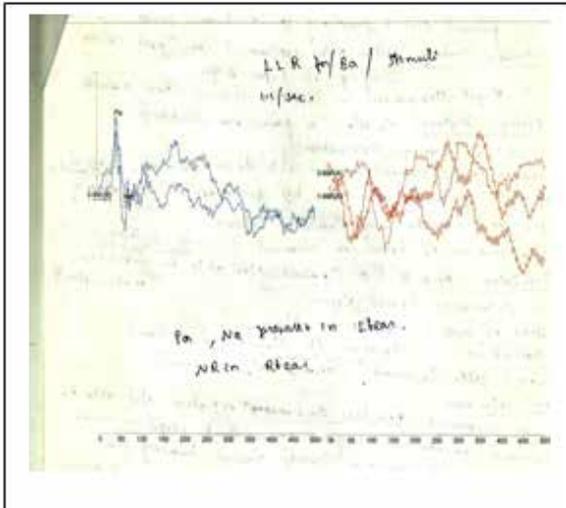
Speech and language assessment revealed severe impairment in auditory comprehension with relatively preserved naming, fluency, reading and writing. Minimal impairment was observed in spontaneous speech and repetition

with occasional phonemic substitutions. Articulation and intelligibility of speech was normal in spontaneous, automated and repetition tasks. No signs of apraxia and dysarthria of speech were observed. He was able to comprehend using visual stimuli like gestures and written modality only. Speech and language evaluation showed no evidence of aphasia.

**Figure 1**



**Figure 1** shows Auditory Brainstem Responses & Figure 2 auditory evoked cortical responses (MLR & LLR) recorded in Right ear (Red tracing) and Left ear (Blue tracing).

**Figure 2****Conclusion**

Electrophysiological tests serve as useful tool in the assessment, diagnosis of central auditory disorders & help in further management of this disorder. Management interventions like the use of hearing aids are not beneficial for this disorder though the patient may present with SNHL. Supportive counseling, speech and language therapeutic interventions like speech reading and auditory stimulation approaches would be beneficial for improving communication and enable independent living of the individual with this disorder.

**Discussion**

Patients with bilateral lesions in the cortical auditory regions manifest with a spectrum of disorders ranging from cortically deaf to generalized auditory agnosia, pure word deafness and/or amusia. In cases of stroke, site of lesion for cortical deafness is typically the middle cerebral artery or its branches bilaterally with spared of auditory brainstem structures [2]. In our audiological evaluation though the puretone Audiometry revealed bilateral moderate SNHL, normal immittance and normal auditory brainstem responses ruled out peripheral and brainstem involvement as a cause for SNHL. In addition to the auditory brainstem responses cortical evoked responses like MLR & LLR were useful for the diagnosis of cortical deafness in this case study. The result of AEP correlates with several previous case reports in the literature on cortical deafness [1, 2 &4]. Also speech and language evaluation helped us in differentiating cortical deafness from aphasia.

**REFERENCE**

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