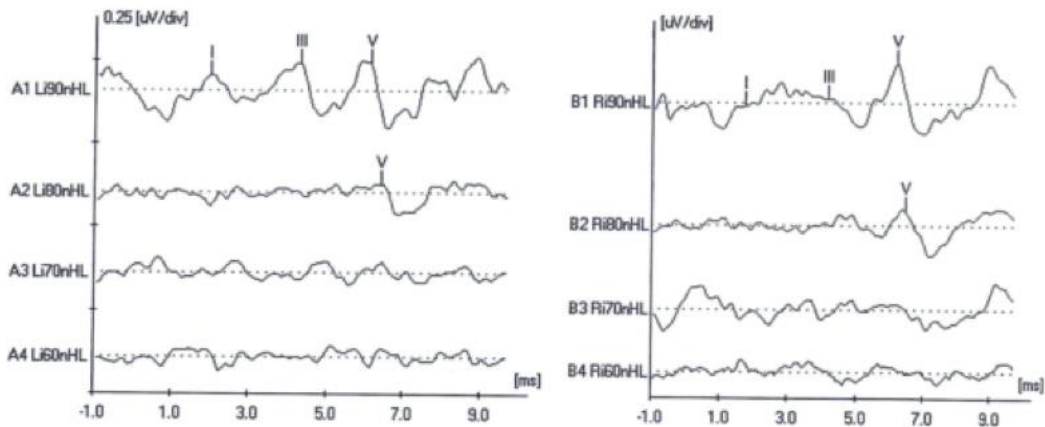
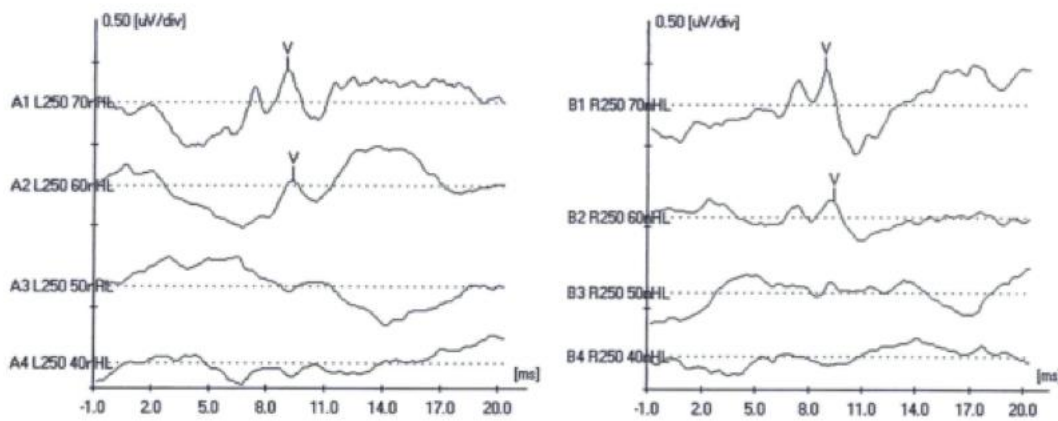


## A. Click-evoked potential ABR



## B. Tone burst ABR



**Figure S1. Analysis of auditory brainstem evoked potentials in proband III3.** (A) Click-evoked potential ABR. The ABR were performed when the subject was aged 4. For both ears, waves I, III, and V were present at the starting intensity of 90 dB nHL. Wave V was replicable at 80 dB nHL but disappeared at 70 dB nHL. (B) Toneburst ABR. A 450-500 Hz toneburst was performed using a Blackman ramp and rarefaction polarity.