

# HEARING ISSUES THAT GO BEYOND THE AUDIOGRAM

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## ABSTRACT

The audiogram has historically been the best predictor of hearing handicap of any single measure used by audiologists. Increasingly, we are finding that the audiogram is a good beginning but does not tell the entire story relative to hearing disorders or hearing handicap. We present illustrative cases representing auditory disorders for which the audiogram by itself is inadequate as a clinical indicator.

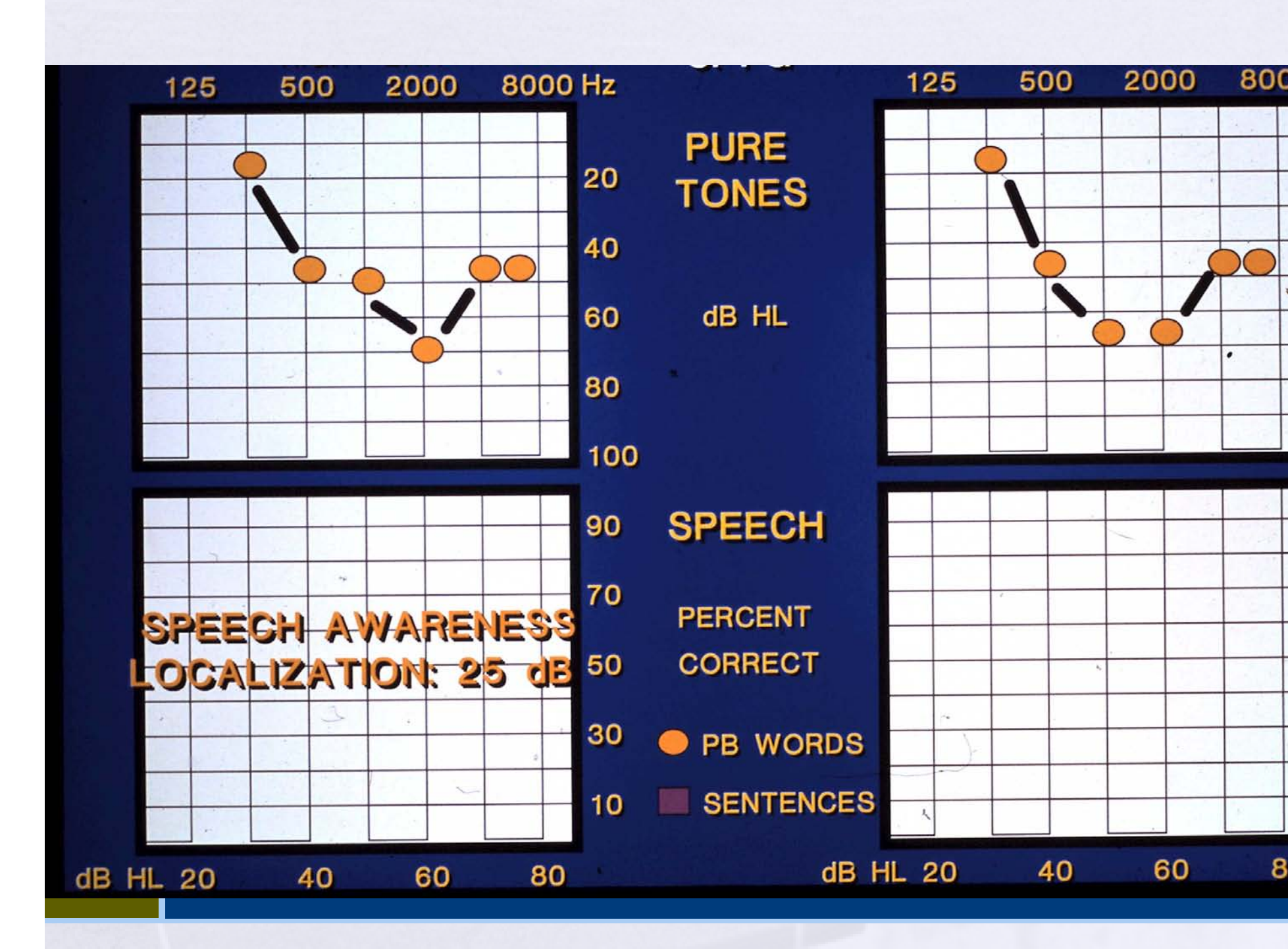
## HIV

Relatively little has been reported concerning HIV and auditory function in children. In a pilot study, we found significant changes in hearing from test to re-test in addition to auditory processing abnormalities, abnormal auditory brainstem responses, and abnormal auditory middle latency responses. These findings occurred in quasi-random fashion and appeared to be independent, unrelated phenomena. They also fluctuated in their occurrence and severity from one visit to another.

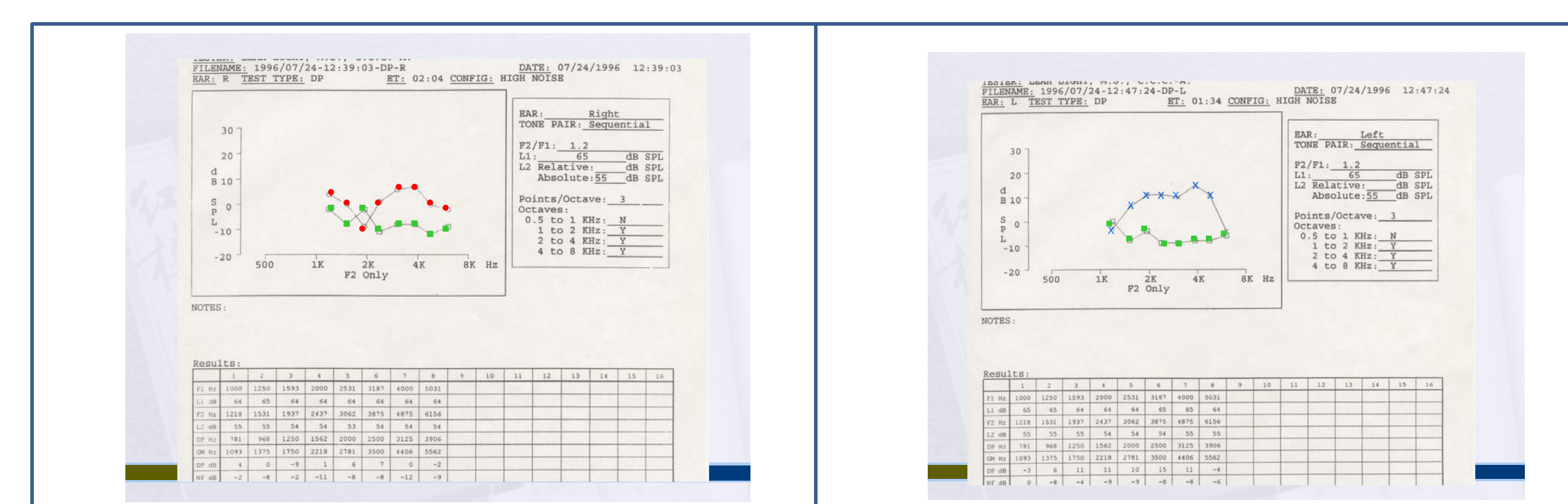
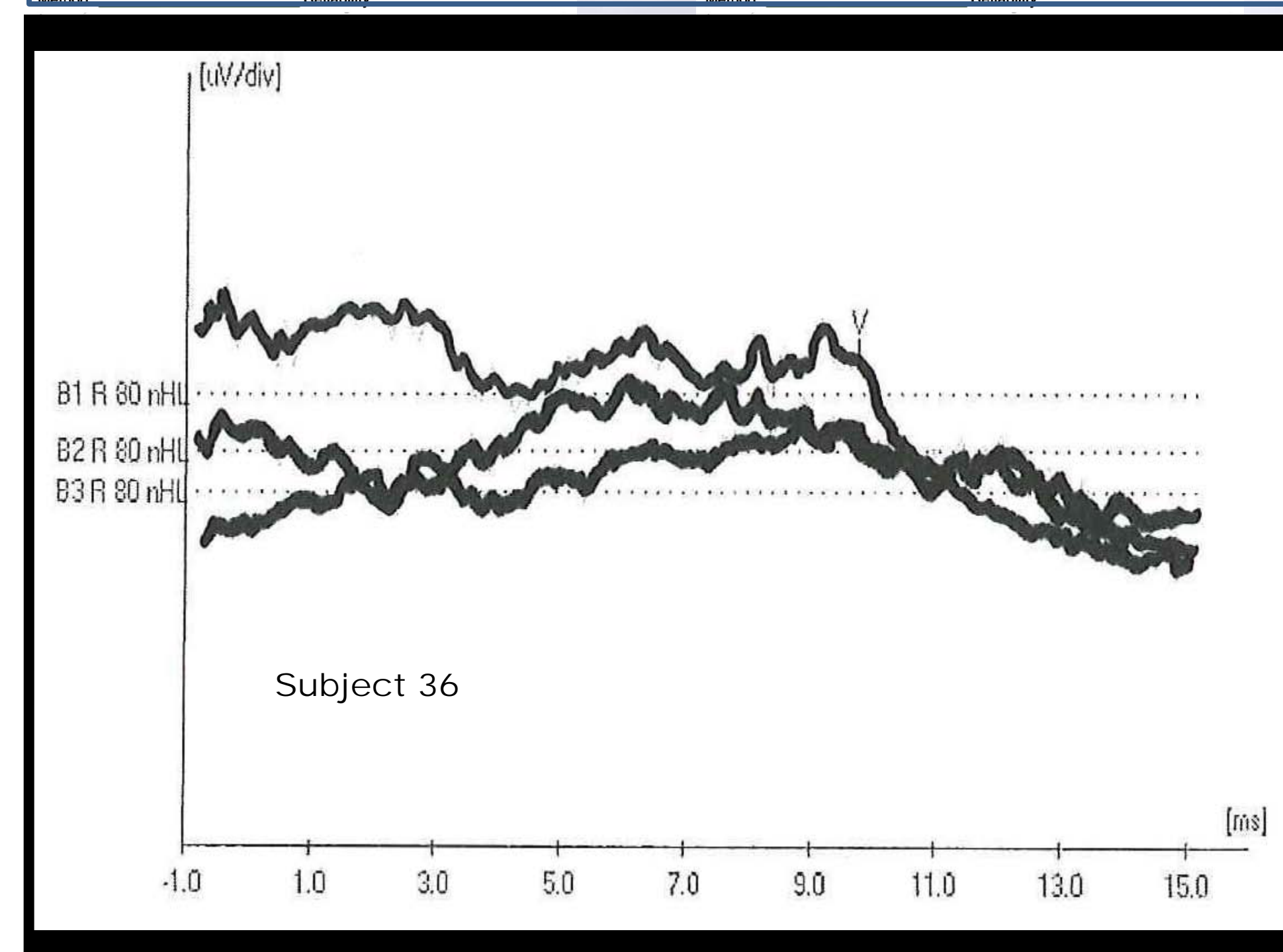
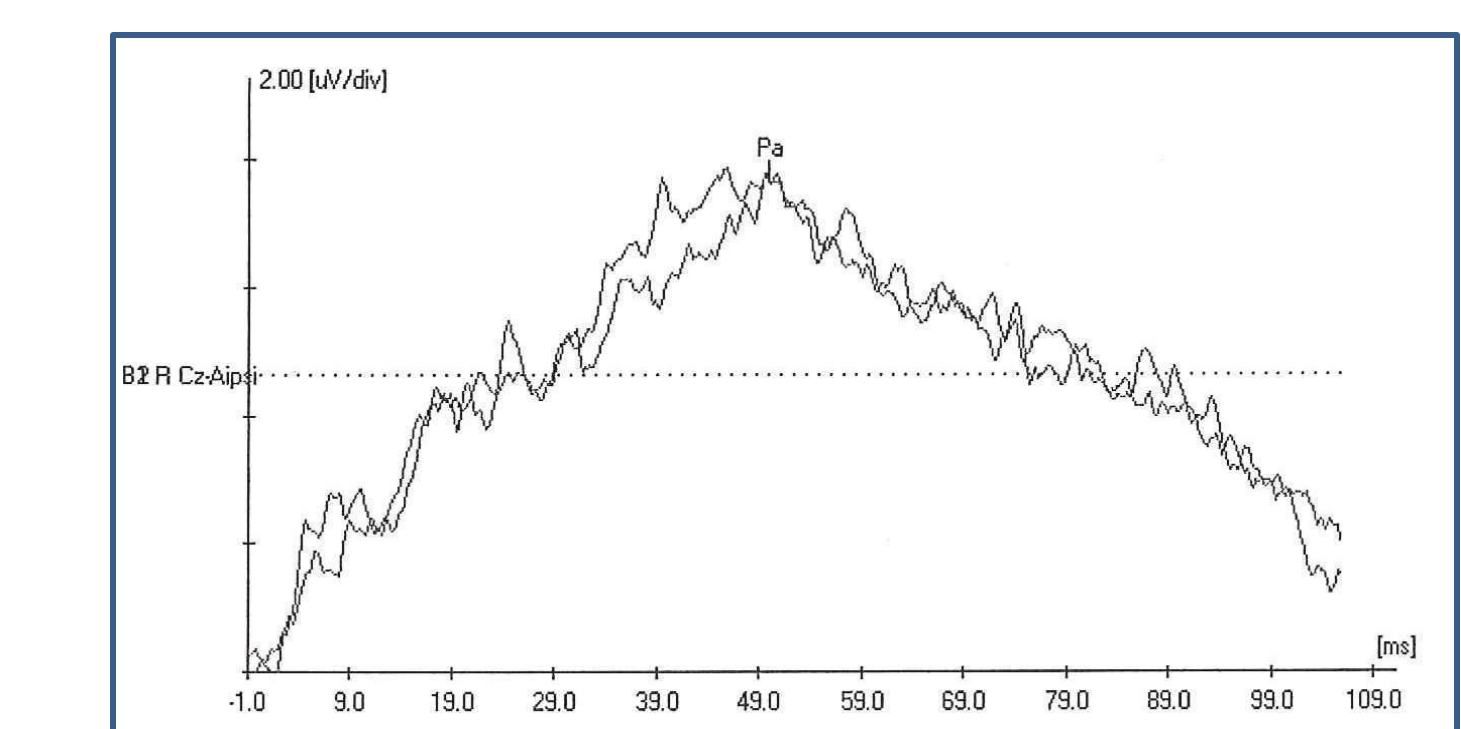
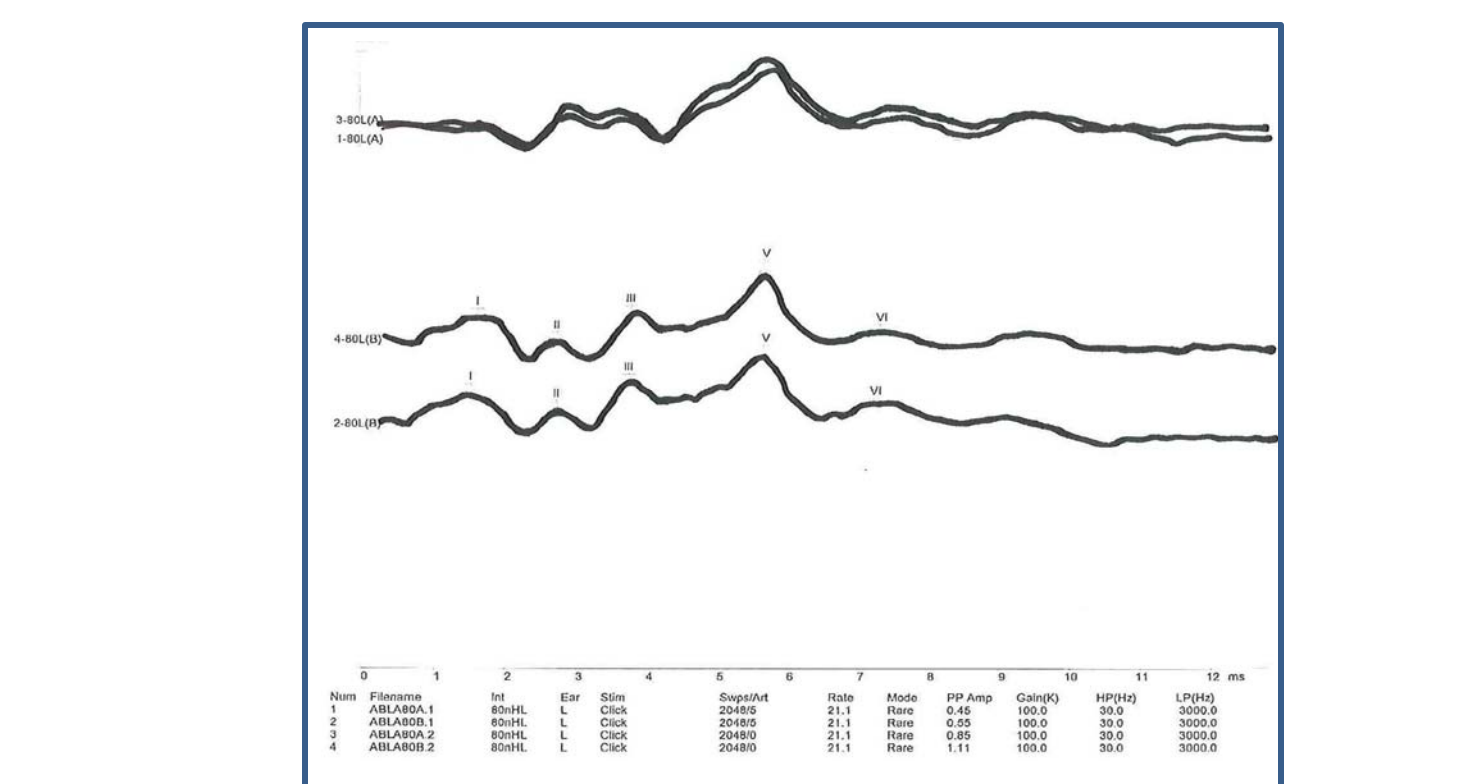
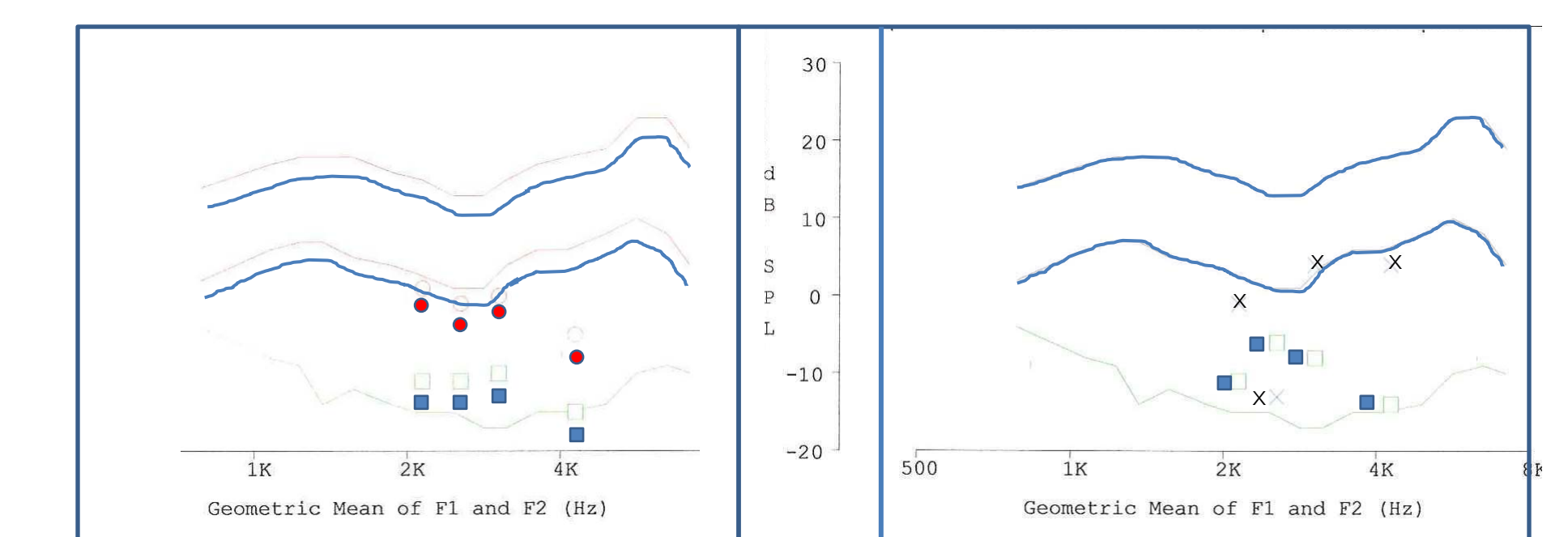
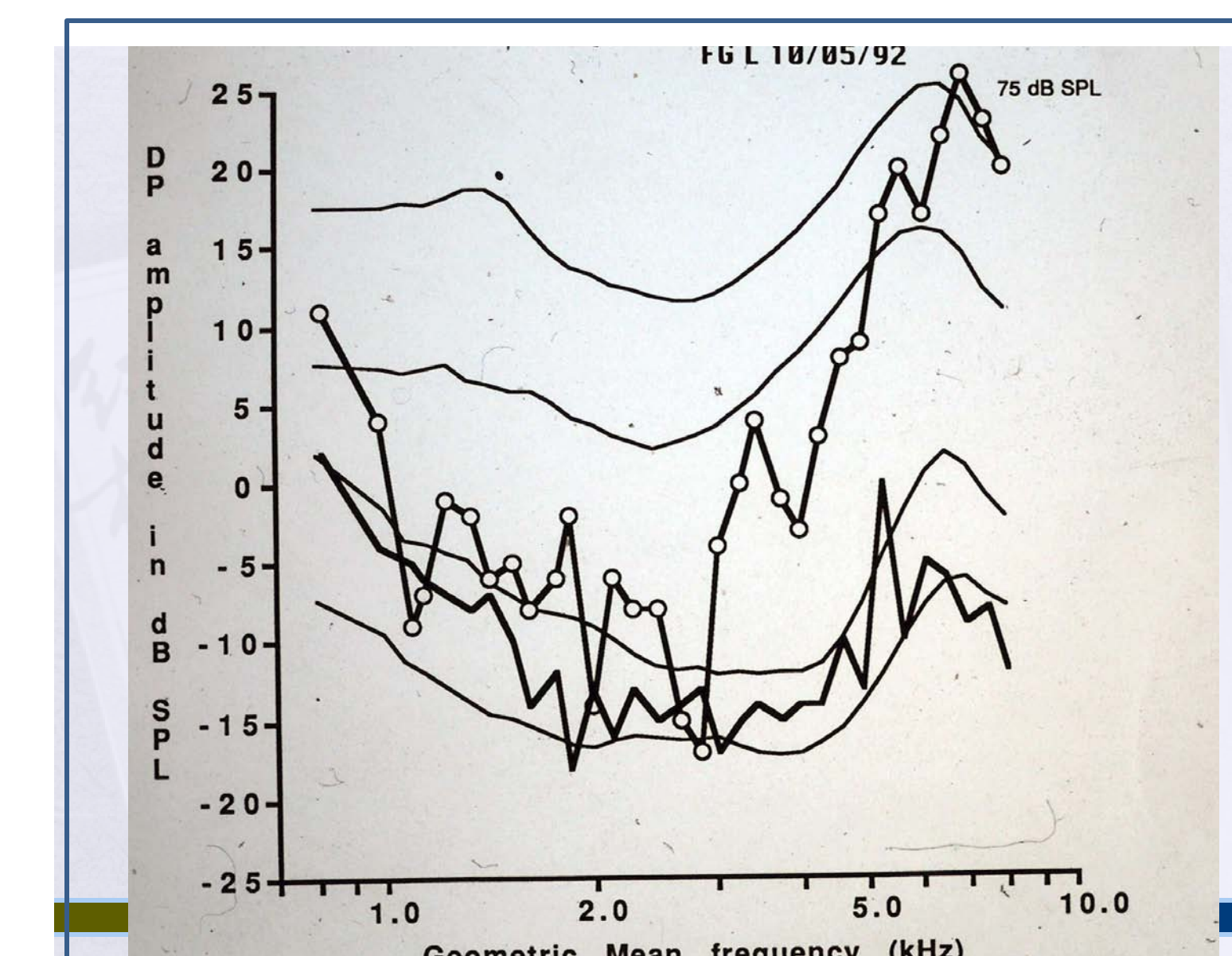
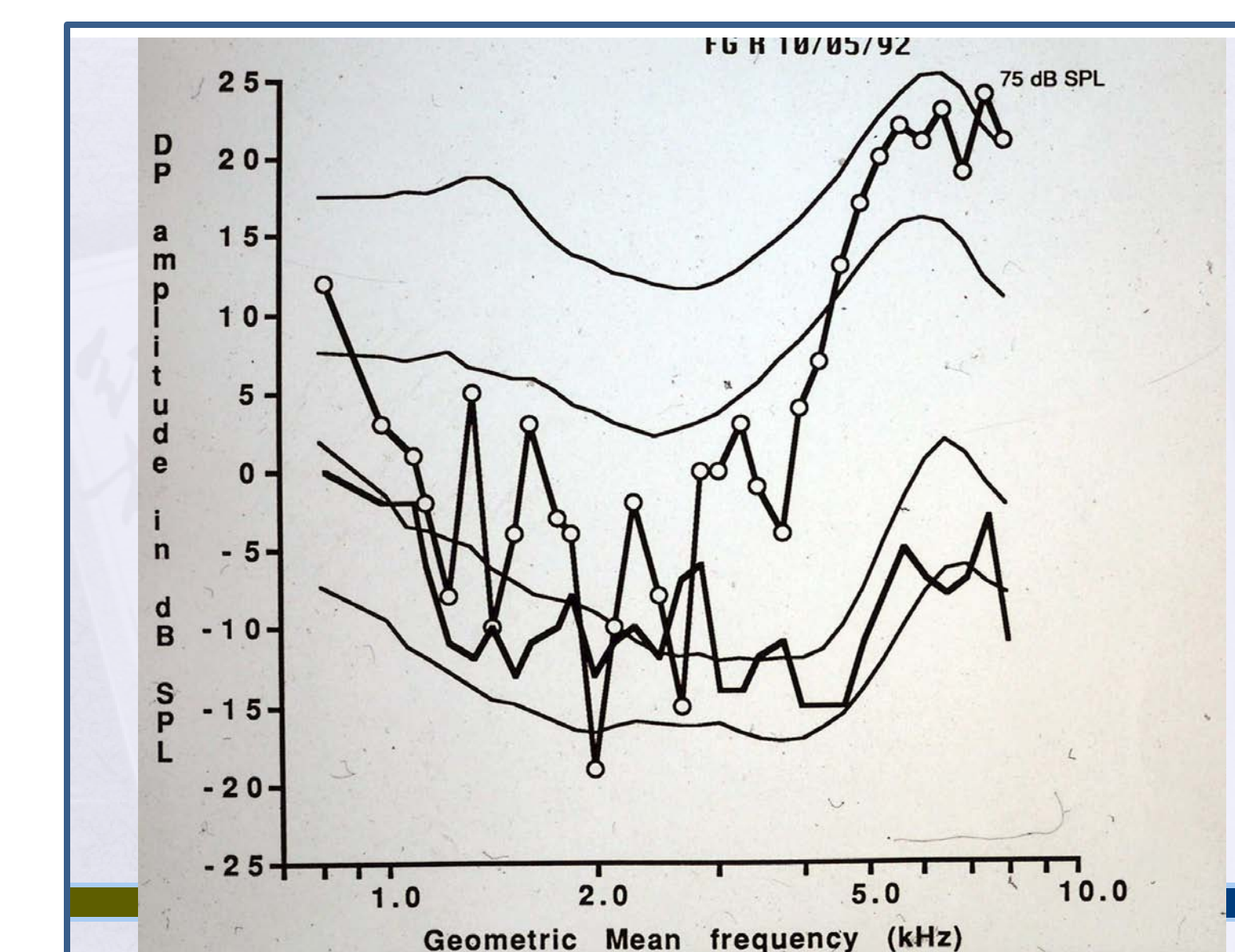
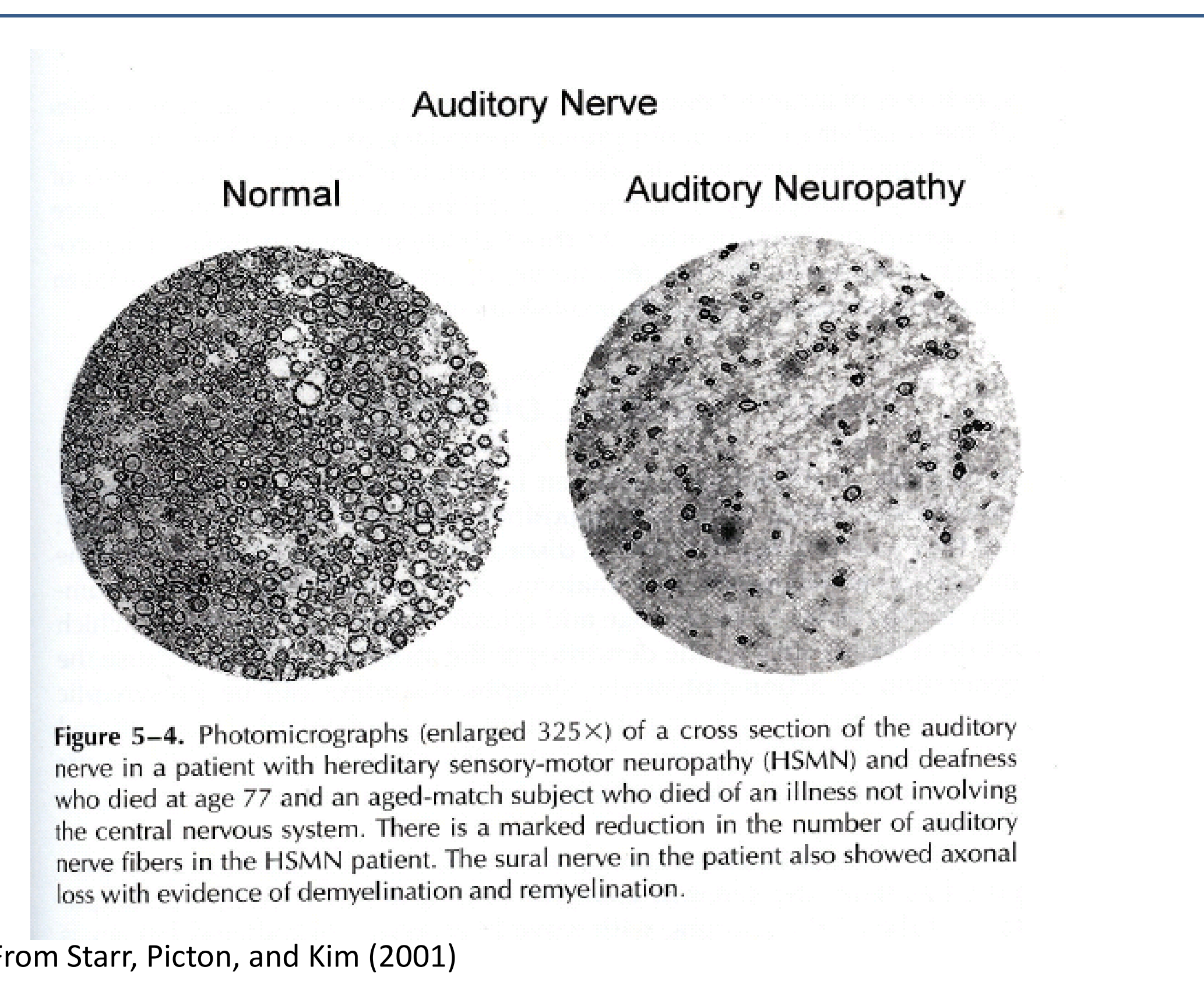
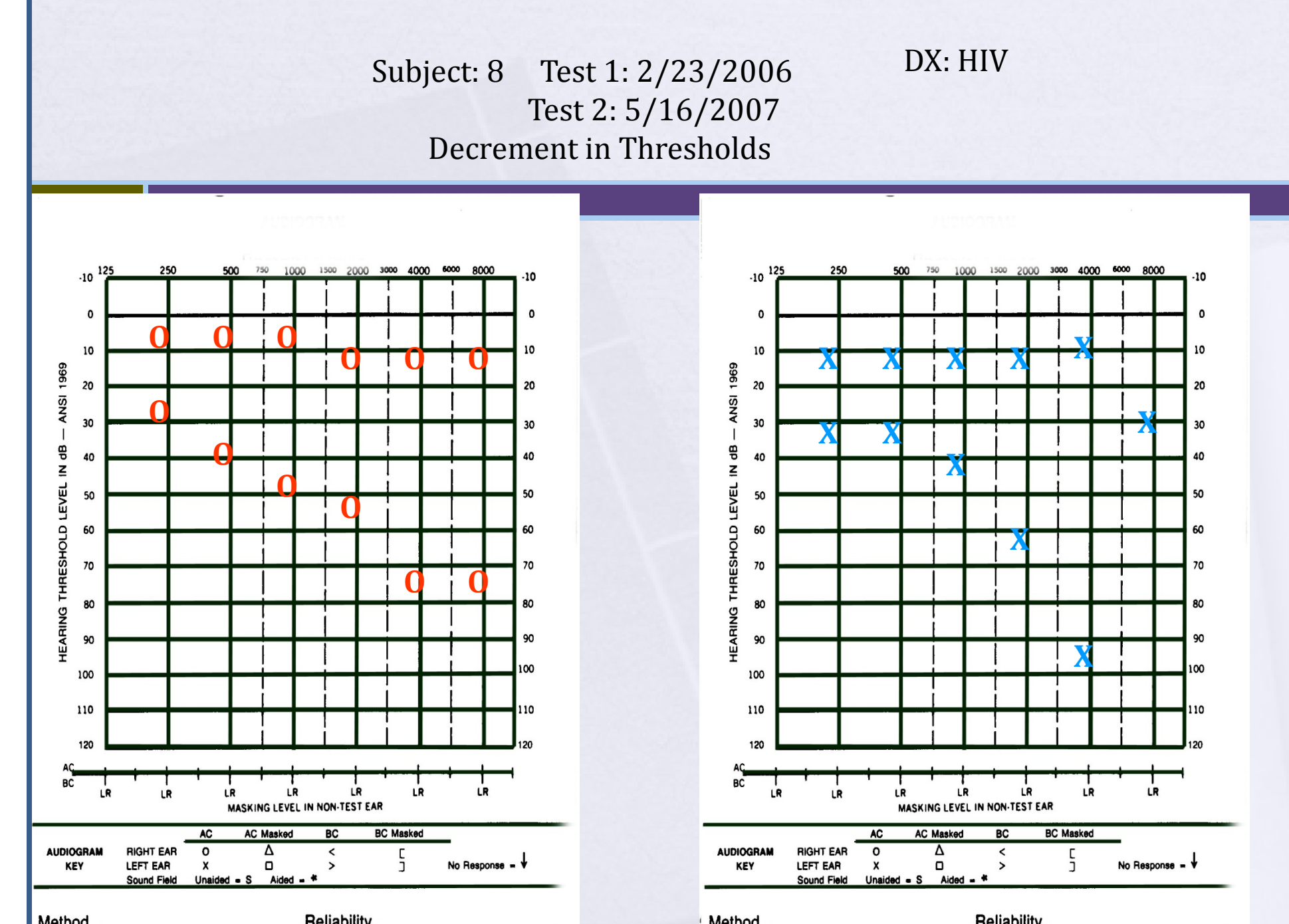
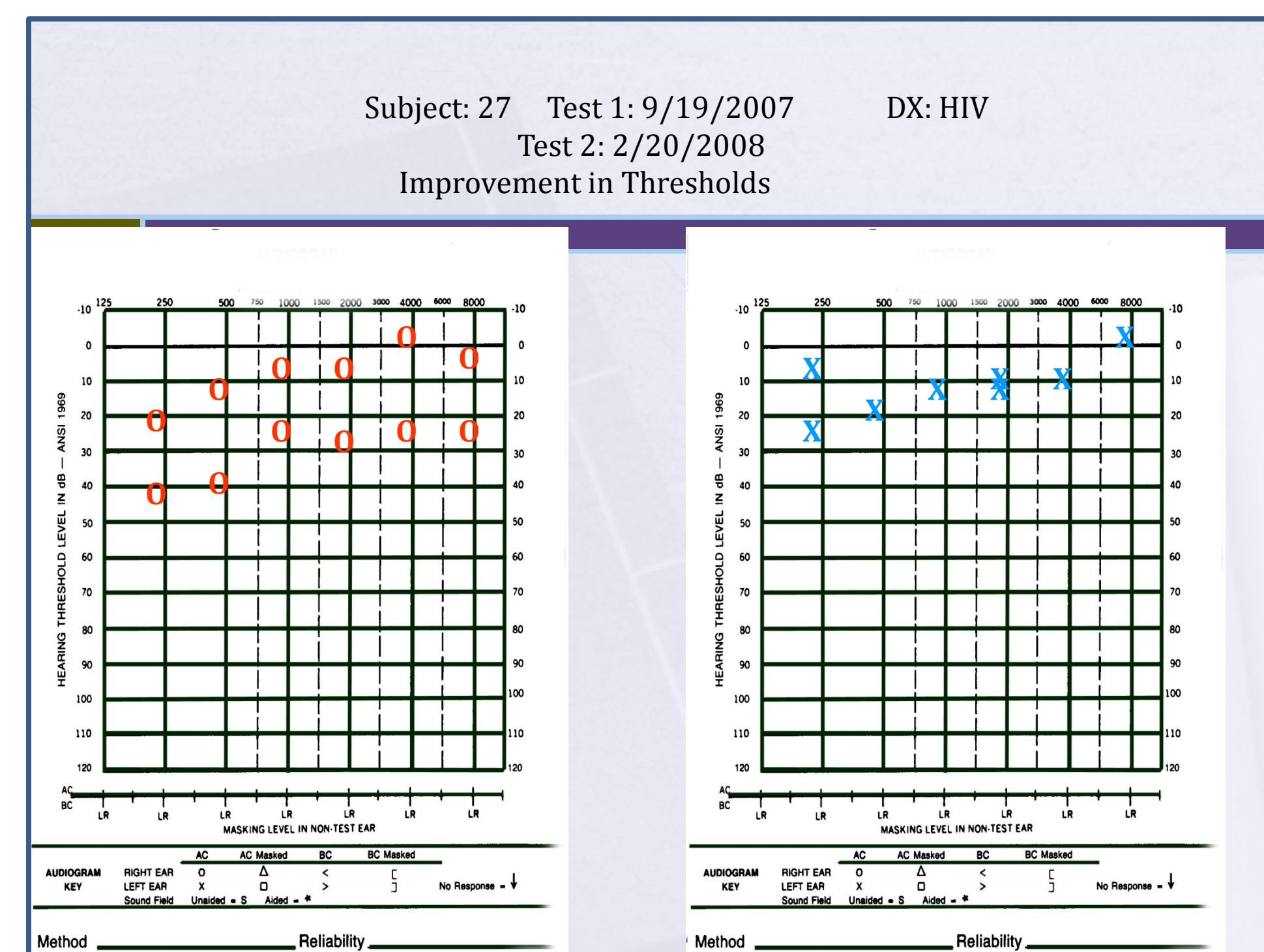
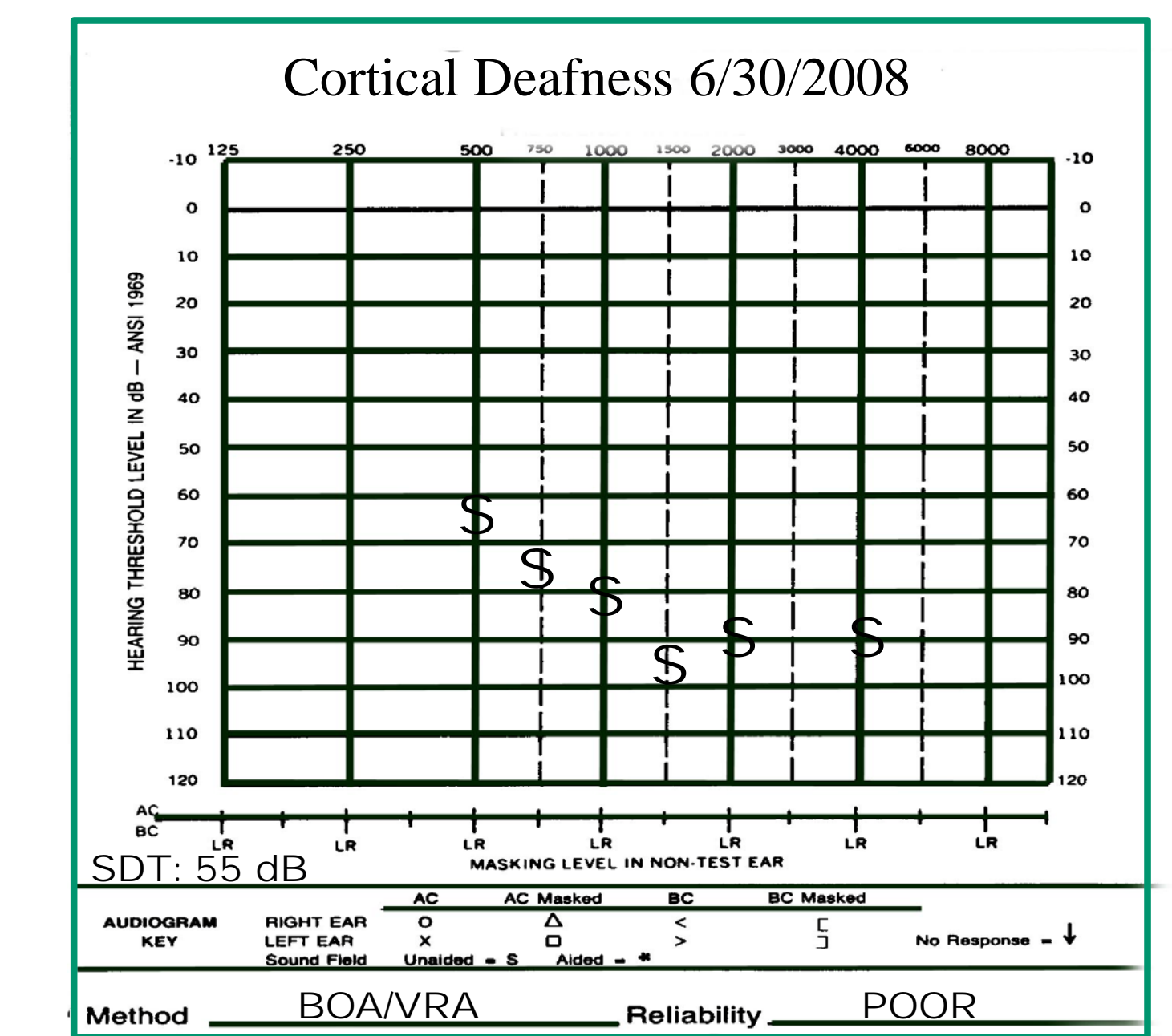
## Auditory Neuropathy / Auditory Dyssynchrony

This disorder disrupts the temporal characteristics of the flow of auditory information between the cochlea and the primary language centers of the brain. It classically has been associated with perinatal hypoxia, hyperbilirubinemia, acidosis, and prematurity. Evidence now exists of a genetic factor of late childhood onset or adult onset, viral infections, immune disorders, fever, or infectious diseases (e.g., parotitis). The associated hearing loss may be low frequency, mild to moderate, or severe. Commonly, there are apparent fluctuations in hearing sensitivity noted from test to re-test. Children with audiograms consistent with severe to profound hearing loss often display reactions to reasonably soft sounds without hearing aids. Even with mild hearing losses, speech understanding is very poor in quiet and especially in background noise. The auditory brainstem response is often absent or highly abnormal. Otoacoustic emissions are often normal, and hearing aids are of little benefit. Children who receive cochlear implants tend not to experience the level of success as children who have hearing loss from other causes. Peripheral neuropathy can be seen similar to that caused by diabetes. The name came from the observation that several early patients had other peripheral neuropathies apart from the auditory nerve.

## Auditory Neuropathy / Auditory Dyssynchrony



## Cortical Deafness



Year of birth: 1996  
History: respiratory distress syndrome, sepsis, hyperbilirubinemia  
Has never been able to use a hearing aid well

Birth at 26 weeks G.A.  
Severe hypoxia and hyperbilirubinemia  
Combined AN / AS with islands of residual hearing

## Discussion

In each of these illustrative cases, the audiogram indicated an auditory problem but failed to reveal the full extent or functional severity of the hearing problem. In the case of HIV, evidence exists to implicate the preference of the virus to congregate in the brainstem. It is quite plausible that the physical impact of viral activity may be manifest by inflammation affecting the brainstem auditory pathways in a manner similar to multiple sclerosis. Certainly, hearing fluctuations indicate a dynamic situation without necessarily producing permanent degradation of the cochlear structures. The abnormal auditory brainstem response points to the central nervous system structures as primary sites of involvement in the total process.

Auditory neuropathy/dyssynchrony occurs most commonly in infants, but onset has occurred at least to 60 years of age. Approximately 27% of patients have no family or medical history to define a cause and effect. The disorder appears to affect both ears in 96% of the reported cases, which means that 4% can display neuropathy symptoms in one ear only. The slide from Starr et al., 2001 indicates that the disease process involves reduction or elimination of the myelin sheath surrounding the nerve fibers disrupting conduction and organization of sensory processing. Valid incidence/prevalence studies for the United States do not exist at this time, but surveys from other countries suggest that the prevalence of 8% to 15% of all hearing impaired children. Effective intervention for these individuals is challenging due to the neural transmission disruption associated with auditory neuropathy. They have disproportionately greater difficulty understanding speech and do not respond as well to hearing aids or cochlear implants as children or adults with more traditional cochlear forms of hearing loss.

Lastly, cortical deafness represents a blockage of information transfer to the primary auditory areas in the cortex. They respond only sporadically to environmental sounds and more readily to music (because elements of music are processed in other areas of the brain). They tend not to respond to speech and verbal communication. Audiologically, the audiogram many times shows partial or complete hearing loss but is in disagreement with outcomes of normal otoacoustic emissions and normal brainstem evoked potentials. For the case shown here, the auditory middle latency response is not normal signifying a problem at the cortical levels in agreement with the other clinical signs and symptoms. These individuals cannot use hearing aids or cochlear implants because the peripheral structures are normal. Most often, the visual pathways and visual language areas are unaffected opening the possibility of communication through sign language.

The primary theme of this presentation focuses on the need to go well beyond the audiogram as a single diagnostic entity when the history, the strength of complaints and magnitude of difficulties, or the diagnostic findings suggest more than a "traditional", straightforward hearing loss. These types of disorders do not occur with great frequency which means that the audiologist cannot be lulled into complacency in performing these evaluations. It is only through diligence and implementation of additional testing when needed that an accurate diagnosis of auditory function can be established and an appropriate plan of intervention developed.