Introduction

Neurodegenerative disorders of childhood encompass a large number of heterogeneous diseases that result from specific genetic and biochemical defects, and a significant group of conditions of unknown cause. Most are inherited in an autosomal recessive fashion and show involvement of different parts of the central and sometimes peripheral nervous system with a variety of progressive deficits, especially dementia, epilepsy, blindness, ataxia and disorders of tone and reflexes ***(Brett and Lake, 1997).***

Neurologic evaluation of pediatric patients is one of the most challenging problems in clinical medicine. Too often, limited cooperation hinders the performance of the neurological physical examination, thus demanding reliance on more objective laboratory measures.

Evoked potentials are very useful clinical tools and its clinical value is fourfold:

- Demonstration of abnormal sensory system function when the history and neurologic examination are equivocal.

- Disclosure of clinically unsuspected malfunction in a sensory system when symptoms or signs in another area of the central nervous system (CNS) suggest demyelinating disease.

- Aid in definition of the anatomic distribution of a disease process.

- Objective monitoring of changes over time in a patient's status.

Thus they provide sensitive, primarily quantitative, extension of the clinical neurologic examination ***(Chiappa et al., 1997).***

Multimodality evoked responses provide more information regarding the functional integrity of several afferent systems in their course from peripheral nerves, through spinal cord and brain stem to the cerebral cortex. These findings when correlated with the results of other conventional tests as EMG, nerve conduction velocities, EEG and CT scan are helpful in the diagnosis and total functional assessment of the nervous system in patients suspected to have degenerative neurological disorders ***(Markand et al., 1982).***

Whereas evoked potentials are useful reliable tools to measure the function of specific parts of the CNS, brain radiography (CT, MRI) reflects morphological abnormalities. CT and MRI are of value in differential diagnosis of neurodegenerative brain disease ***(Vanhanen et al., 1994).***