Hereditary Progressive Dystonia With Marked Diurnal Fluctuation

This study examines the condition known as parkinsonism of early onset with diurnal fluctuation (PEDF). It aims to clarify the differences between PEDF and early-onset Parkinson disease, then to compare PEDF with HPD (hereditary progressive dystonia with marked diurnal fluctuation). For the purpose of long-term observation of PEDF, previously observed patients were re-examined after 17-23 years. Of 11 patients from four families, one patient had already died at the time of the previous study and subsequently two others died. Eight patients, therefore, were examined in the present study. Brain computerized tomography (CT), magnetic resonance imaging (MRI) and single photon emission CT (SPECT) with 125-imipramine were performed on some of the patients. To differentiate PEDF from early-onset Parkinson disease, the investigators collected data for 40 cases of parkinsonism with onset before the age of 40, including the above referred cases from cumulative case records. All patients were thoroughly examined to exclude those with any known cause of secondary parkinsonism (Huntington disease and so on). The 40 cases were divided into two groups according to the presence or absence of diurnal fluctuation, and differences between the two in familial occurrence, sex ratio, age at onset and neurological manifestations were examined.

Adv Neurol. 198745:227-34. Hereditary progressive dystonia with marked diurnal fluctuation: clinicopathophysiological identification in reference to juvenileAbstract. The causative gene for hereditary progressive dystonia with marked diurnal fluctuation/dopa-responsive dystonia (HPD/DRD) was discovered in 1994Hereditary progressive dystonia with marked diurnal fluctuation--consideration on its pathophysiology based on the characteristics of clinical andBrain Dev. 2000 Sep22 Suppl 1:S102-6. Gene mutation in hereditary progressive dystonia with marked diurnal fluctuation (HPD), strictly definedSummary. Hereditary progressive dystonia with marked diurnal fluctuation is a particular levodopa-responsive dystonia with onset in childhood. Clinically, it isAbstract. Hereditary progressive dystonia with marked diurnal fluctuation (HPD) is a childhood-onset, postural dystonia that is characterized by marked diurnalHereditary progressive dystonia with marked diurnal fluctuation or the strictly defined dopa-responsive dystonia (HPD/DRD) is an autosomally dominantlyHereditary progressive dystonia with marked diurnal fluctuation (HPD) is an

autosomally dominantly inherited postural dystonia with onset in childhood which is.Since 1988, we have diagnosed 6 cases of hereditary progressive dystonia with marked diurnal fluctuation (HPD) in Taiwan. All cases presented with clinicalAutosomal dominant dopa-responsive dystonia (DYT5a) is a rare marked diurnal fluctuation Hereditary progressive dystonia with marked diurnal fluctuation.Long-term effects of l-dopa on hereditary progressive dystonia with marked diurnal fluctuation. In: Berardelli A et al., editors. Motor disturbances II. London: Hereditary progressive dystonia with marked diurnal fluctuation (HPD) (also known as dopa responsive dystonia) is a dystonia with onset inBrain Dev. 1994 Mar-Apr16(2):126-31. Hereditary progressive dystonia with marked diurnal fluctuation (Segawa syndrome) in Taiwan. Wang PJ(1), Ko YM,Hereditary progressive dystonia with marked diurnal fluctuation (HPD) is a dopa-responsive dystonia, now called autosomal dominant GTP cyclohydrolase 1