# Pediatric Neurology Part III: Chapter 178. Progressive myoclonus epilepsy (Handbook of Clinical Neurology)

*Jean-Marie Girard, Julie Turnbull, Nivetha Ramachandran, Berge A. Minassian*

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The progressive myoclonus epilepsies (PMEs) consist of a group of diseases with myoclonic seizures and progressive neurodegeneration, with onset in childhood and/or adolescence. Lafora disease is a neuronal glycogenosis in which normal glycogen is transformed into starch-like polyglucosans that accumulate in the neuronal somatodendritic compartment. It is caused by defects of two genes of yet unknown function, one encoding a glycogen phosphatase (laforin) and the other an ubiquitin E3 ligase (malin). Early cognitive deterioration, visual seizures affecting over half, and slowing down of EEG basic activity are three major diagnostic clues. Unverricht–Lundborg disease is presently thought to be due to damage to neurons by lysosomal cathepsins and reactive oxygen species due to absence of cystatin B, a small protein that inactivates cathepsins and, by ways yet unknown, quenches damaging redox compounds. Preserved cognition and background EEG activity, action myoclonus early morning and vertex spikes in REM sleep are the diagnostic clues. Sialidosis, with cherry-red spot, neuronopathic Gaucher disease, with paralysis of verticality, and ataxia-PME, with ataxia at onset in the middle of the first decade, are also lysosomal diseases. How the lysosomal defect culminates in myoclonus and epilepsy in these conditions remains unknown.

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