

# An Update to Epilepsy Syndromes with a Variable Age of Onset

The ILAE Task Force on Nosology and Definitions provides guidelines for classifying epilepsy syndromes with a variable age of onset

## Background

A considerable number of epilepsy syndromes present at variable ages. However, these syndromes are not as well-recognised. To aid with syndrome identification and disease management, the International League Against Epilepsy (ILAE) Task Force has updated their guidelines on the classification of epilepsy syndromes that present later in life, with a variable age of onset.

## What this study addresses

This position paper delineates the diagnostic criteria—including specific clinical features, indicators, and phenotypes—of various epilepsy syndromes showing onset at a variable age, while also providing standardised definitions for the same.

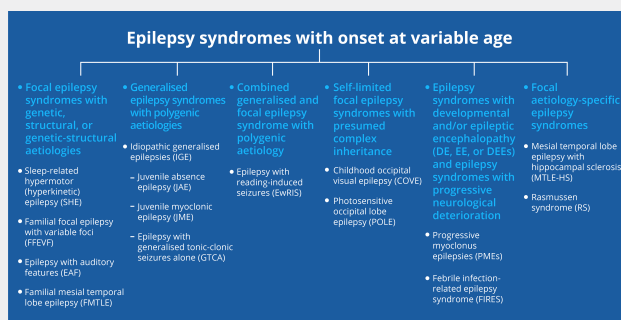
## Main findings

Based on seizure presentation, EEG and imaging patterns, evolution of the disease course, and other characteristics, these syndromes can be classified in the following groups: generalised epilepsy syndromes, self-limited focal epilepsy syndromes, focal epilepsy syndromes, combined generalised and focal epilepsy syndromes, epilepsy syndromes with developmental and/or epileptic encephalopathies (DE, EE, or DEEs) and epilepsy syndromes with progressive neurological deterioration, and lastly, Aetiology-specific epilepsy syndromes. The guidelines also redefine syndrome nomenclature to reflect the main electroclinical phenotypes (such as mandatory seizure type) and aetiology, while veering away from eponymous names.

## Key takeaway

Syndrome identification often has important treatment and prognostic implications; therefore, the updated guidelines could bolster improvements in disease management and treatment outcomes in epilepsy syndromes that present at a variable age.

**Figure: Classification and updated nomenclature of epilepsy syndromes that present at an older, variable age**



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