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Redefining Childhood-Onset Epilepsy Syndromes

Updated classification and clinical management guidelines

Epilepsy is a neurological disorder associated with abnormal electrical activity in the brain, which causes sudden recurrent episodes of sensory disturbance, loss of consciousness, or convulsions

For efficient management of epilepsy in children, diagnosis of their exact syndrome is critical

This requires:

Defining specific clinical and laboratory features of childhood-onset epilepsy syndromes

Using terms that clearly express clinical features of each syndrome

As the clinical understanding of epilepsy is evolving continuously, for accurate diagnosis, determination of prognosis, and management of this condition, updating the 2017 Epilepsy and Seizure classification given by ILAE is essential

The most recent:
Classification | Clinical features | Laboratory features 〉 Of epilepsy syndromes that present in childhood are thus summarised here

Typically, epileptic syndromes are diagnosed by analysing the:

- Clinical presentation of seizures
- Disease's evolution over time
- Child's developmental progress
- EEG features
- Case-specific brain magnetic resonance imaging (MRI)

Childhood-onset epilepsy syndromes

Self-limited focal epilepsies

- Self-limited epilepsy with centrotemporal spikes
- Self-limited epilepsy with autonomic seizures
- Childhood occipital visual epilepsy
- Photosensitive occipital lobe epilepsy

Genetic generalised epilepsies

- Childhood absence epilepsy
- Epilepsy with myoclonic absence
- Epilepsy with eyelid myoclonia

Developmental and/or epileptic encephalopathies

- Epilepsy with myoclonic-atonic seizures
- Lennox-Gastaut syndrome
- Developmental and/or epileptic encephalopathy with spike–and–wave activation in sleep
- Hemiconvulsion–hemiplegia–epilepsy syndrome
- Febrile infection-related epilepsy syndrome

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Self-limited focal epilepsies (SeLFEs)

- Focal epilepsy syndromes, with childhood-onset
- SeLFEs account for up to 25% of all childhood epilepsies

General characteristics:

- Age-dependent occurrence
- Responsive to anti-epileptic medicines
- Normal cognition and neurological examination
- No significantly associated brain lesion
- Classical seizure semiology for each syndrome
- Remission usually occurs by puberty
- A lack of disease history
- Specific EEG features

Clinical characteristics and diagnosis of SeLFEs

Based on their long-term prognosis, SeLFEs are divided into two subgroups:

Subgroup 1: Remission expected by the time of adolescence; treatment to be stopped by then

Subgroup 2: Most patients experience remission, but some may continue experiencing seizures post-adolescence, requiring chronic treatment with anti-seizure medications

	Age of onset	Seizures	EEG	Neurological exam	Requirements for diagnosis
Self-limited epilepsy with autonomic seizures (SeLEAS)	Range: 1-14 years Peak: 3-6 years	• Focal seizures • With autonomic symptoms like retching and vomiting, malaise, pallor, flushing, abdominal pain, and pupillary or cardio-respiratory changes	• High amplitude, focal or multifocal high voltage epileptiform abnormalities, which increase in drowsiness and sleep		• Interictal EEG
Self-limited epilepsy with centrotemporal spikes (SeLECTS)	Range: 4-10 years Peak: 7 years	• Focal seizures • With dysarthria, sialorrhea, dysphasia, and unilateral clonic or tonic-clonic movement of mouth • Nocturnal focal to bilateral tonic-clonic seizures in sleep only • If seizures occur during sleep, they are seen within 1-2 hours of falling asleep or 1-2 hours prior to awakening	• High amplitude, centrotemporal biphasic epileptiform abnormalities	• Hemiparesis or focal neurological findings	
Childhood occipital visual epilepsy (COVE)	Range: 4-7 years Mean: 11 years	• Focal sensory visual seizures - With visual phenomena - Sometimes, with motor signs • Seizures arise mostly on waking up	• Occipital spikes or spike–and–wave abnormalities (awake or sleep)		• Interictal EEG • MRI to rule out brain lesions
Photosensitive occipital lobe epilepsy (POLE)	Range: 15 months 19 years Peak: 8-9 years	• Focal sensory visual seizures • May evolve to bilateral tonic-clonic seizures • Triggered by optic stimuli, like flickering lights	• Occipital epileptiform abnormalities facilitated by eye closure and intermittent photic stimulation (IPS)		• Ictal EEG • MRI to rule out brain lesions

Genetic generalised epilepsies (GGEs)

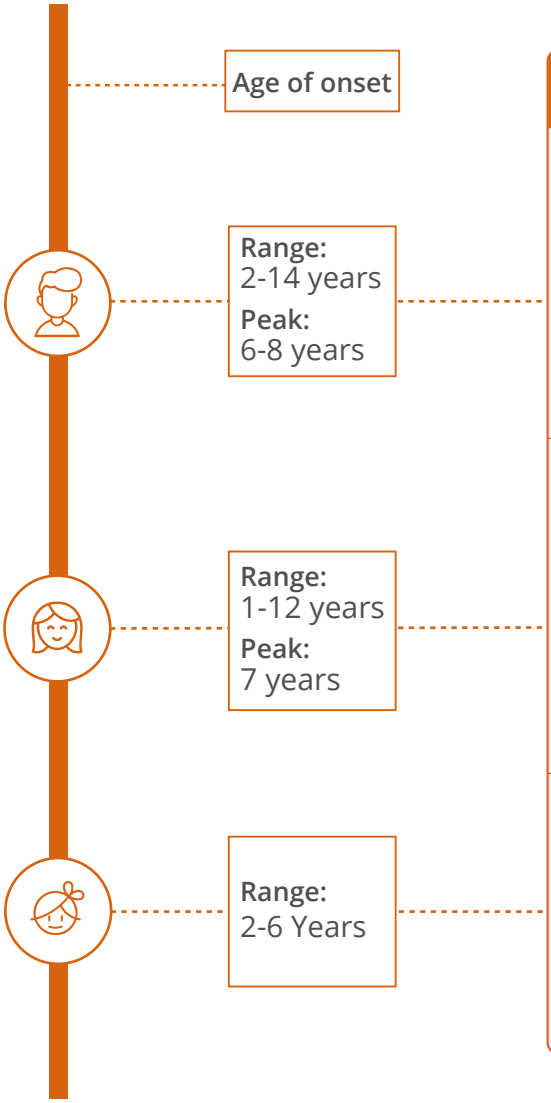
General characteristics:

Positive family history of epilepsy

Variable cognition and neurological examination

May exhibit drug resistance

Clinical characteristics and diagnosis of GGEs



	Seizures	EEG	Requirements for diagnosis
Epilepsy with eyelid myoclonia (E-EM)	• Eyelid myoclonia (short, jerking spasms)	• Eye closure and intermittent photic stimulation elicits fast (3-6 Hz) generalised polyspikes or polyspike–and–slow–wave	• Proof of myoclonia • Interictal EEG
Epilepsy with myoclonic absences (E-MA)	• Myoclonic absence seizures	• Regular 3 Hz generalised spike–and–slow–wave time-locked with the myoclonic jerks	• Clinical features • Consider MRI to rule out any underlying cause
Epilepsy with Myoclonic-Atonic Seizures (EMAtS)	• Myoclonic-atonic seizures	• Generalised 2-6 Hz spike wave or multiple spikes–and–wave	• Clinical features • Consider MRI to rule out any underlying cause

Developmental and epileptic encephalopathies (DEEs)

Clinical characteristics and diagnosis

	Seizures	EEG	Neurological exam	Requirements for diagnosis
Lennox-Gastaut syndrome (LGS)	• Tonic seizures, in combination with any of the following: - Atypical absences - Atonic - Myoclonic - Focal impaired awareness - Generalised tonic clonic - Nonconvulsive status epilepticus - Epileptic spasms	• Generalised slow spike-wave <2.5 Hz (or history of this finding on prior EEG) • Generalised paroxysmal fast activity in sleep (or history of this finding on prior EEG)		• Age generally between 10-18 years • Interictal EEG with specific characteristics • MRI to rule out any underlying aetiology
Developmental and/or epileptic encephalopathy with spike–and–wave activation in sleep (DEE-SWAS and/or EE-SWAS) Landau-Kleffner syndrome, Epileptic Encephalopathy with Continuous Spike–and–Wave in Sleep, Atypical (Benign) Partial Epilepsy (pseudo-Lennox syndrome)	• Infrequent and drug-responsive seizures are observed during the initial phase between 2 and 5 years of age • These early seizures are typically: - Focal motor, with or without impaired awareness - Focal to bilateral tonic-clonic seizures • Seizures typically worsen with the evolution of multiple seizure types	• Slow (1.5-2 Hz) spike–and–wave abnormalities in N-REM sleep • Abnormalities are markedly activated in sleep		• Sleep EEG • Cognitive, behavioural, or motor regression • Plateauing temporally related to SWAS on EEG
Febrile infection-related epilepsy syndrome (FIRES) Acute encephalitis with refractory, repetitive partial seizures (AERRPS), Devastating epileptic encephalopathy in school-aged children (DESC)	• Focal and multifocal seizures • Often evolve to bilateral tonic-clonic seizures • Progress in frequency and severity to culminate in super-refractory status epilepticus typically within 2 weeks of onset	• Slowing of the background activity with multifocal epileptiform abnormalities and frequent, focal electrographic and electroclinical seizures	• Neurological examination abnormalities prior to onset of seizures	• History of fever in the 2 weeks preceding seizure onset • Acute encephalopathy with onset of frequent seizures • MRI and ictal EEG mandatory
Hemiconvulsion–hemiplegia–epilepsy syndrome	• Acute stage: Episode of febrile, hemiclonic status epilepticus immediately followed by permanent hemiparesis • Chronic stage: Unilateral focal motor or focal–to–bilateral tonic–clonic seizures within 3 years of initial status epilepticus	• Slowing of background activity over the affected hemisphere • Focal or multifocal epileptiform abnormalities over the affected hemisphere in the chronic phase	• Focal neurological abnormalities prior to initial episode of febrile status epilepticus • Facial angioma suggestive of Sturge-Weber syndrome	• MRI or computed tomography (CT) scan immediately following the acute phase

Reference:
Specchio, N., Wirrell, E.C., Scheffer, I.E., Nabbout, R., Riney, K., … & Auvin, S. (2022). International League Against Epilepsy classification and definition of epilepsy syndromes with onset in childhood: position paper by the ILAE Task Force on Nosology and Definitions. *Epilepsia,* 1– 45, https://doi.org/10.1111/epi.17241.

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