

EPILEPSY

Definition

a neurological condition characterised by recurrent unprovoked epileptic seizures

An *epileptic seizure* is the clinical manifestation of an abnormal and excessive discharge of a set of neurons in the brain.

An *epileptic syndrome* is a complex of signs and symptoms that define a unique epilepsy condition. Syndromes are classified on the basis of seizure type(s), clinical context, EEG features and neuroimaging

Epilepsy should be viewed as a symptom of an underlying neurological disorder and not as a single disease entity.

Table 1. Differential diagnosis of seizures

Neonates and infants	Young children	Childhood and Adolescents
jitteriness	breath holding spells	vasovagal syncope
benign myoclonus	reflex anoxic seizures	migraine
apnoea	parasomnia	narcolepsy
gastro-oesophageal reflux	benign paroxysmal vertigo	panic attacks
shuddering attacks	paroxysmal choreoathetosis	pseudoseizures
benign paroxysmal torticollis	tics and ritualistic movements	Any Age
hyperekplexia	rage attacks	drug-induced dystonia
		cardiac dysrhythmias

Management and Investigations

- detailed history of the seizures. Also note birth history, developmental milestones, family history.
- look for dysmorphism, neurocutaneous signs; do thorough CNS, developmental examination.
- investigations are recommended when a *second* afebrile seizure occurs:
 - routine biochemical tests only if clinical features suggest a biochemical disorder, e.g. hypoglycaemia or hypocalcaemia
 - ECG may be performed if suspicion of a cardiac dysrhythmia.
 - EEG is important to support the clinical diagnosis of epileptic seizures, classify the epileptic syndrome, selection of anti-epileptic drug and prognosis. It also helps in localization of seizure foci in intractable epilepsy
- neuroimaging (preferably MRI) is indicated for any child with
 - epilepsy occurring in the first year of life, except febrile seizures
 - partial epilepsy except benign rolandic epilepsy
 - developmental delay or regression

Table 2. ILAE Classification of seizure types

Partial
simple
complex
partial seizure with secondary generalisation
Generalised
absence
atypical absences
myoclonic
tonic-clonic
tonic
clonic
atonic
Unclassified
infantile spasms

ILAE = International League Against Epilepsy

Table 3. Classification of epilepsies and epileptic syndromes (ILAE 1989)**Focal or Partial epilepsies & syndromes***Idiopathic*

benign childhood epilepsy with
centrotemporal (Rolandic) spikes
childhood epilepsy with occipital paroxysms
primary reading epilepsy

Symptomatic

epileptia partialis continua
syndrome characterised by specific
modes of precipitation
temporal lobe epilepsy
frontal lobe epilepsy

**Epilepsies and syndromes undetermined,
whether focal or generalised**

neonatal seizures
severe myoclonic epilepsy in infancy
epilepsy with CSWS
(Continuous spike waves of slow sleep)
acquired epileptic aphasia
(Landau-Kleffner syndrome)

Special syndromes; situation-related seizures

febrile convulsions
isolated seizures or isolated status epilepticus
seizures occurring only with an acute
metabolic or toxic event

Generalized epilepsies and syndromes*Idiopathic*

benign neonatal familial convulsions
benign neonatal convulsions
benign myoclonic epilepsy in infancy
childhood absence epilepsy
juvenile absence epilepsy
juvenile myoclonic epilepsy
epilepsy with grand mal seizures
on awakening

Cryptogenic or symptomatic

West syndrome
Lennox-Gastaut syndrome
epilepsy with myoclonic atstatic seizures
epilepsy with myoclonic absences

Symptomatic

early myoclonic encephalopathy
early infantile epileptic encephalopathy
other symptomatic generalised epilepsies
not defined above
specific syndromes

Principles of anticonvulsant therapy for Epilepsy

- treatment recommended ≥ 2 episodes (recurrence risk up to 80%)
- attempt to classify the seizure type(s) and epileptic syndrome. Monotherapy as far as possible. Choose most appropriate drug, increase dose gradually until epilepsy controlled or maximum dose reached or side effects occur.
- add on the second drug if first drug failed. Optimise second drug, then try to withdraw first drug. (alternative monotherapy)
- rational combination therapy (usually 2 or maximum 3 drugs) i.e. combines drugs with different mechanism of action and consider their spectrum of efficacy, drug interactions and adverse effects.
- monitor drug levels (usually with carbamazepine, phenytoin, phenobarbitone) to check compliance, if seizures not well controlled despite adequate doses or in situations of polypharmacy where drug interaction is suspected.
- when withdrawal of medication is planned (generally being seizure free for 2 years), consideration should be given to epilepsy syndrome, likely prognosis and individual circumstances before attempting slow withdrawal of medication over 3-6 months (maybe longer if clonazepam or phenobarbitone). If seizures recur, the last dose reduction is reversed and medical advice sought.

Table 4. Selecting anticonvulsants according to seizure type

seizure type	first line	second line
Partial seizures		
simple partial	carbamazepine	lamotrigine
complex partial	valproate	topiramate
secondarily generalised		levetiracetam, phenytoin phenobarbitone, clonazepam
Generalised seizures		
tonic-clonic	valproate	lamotrigine
clonic		topiramate, clonazepam carbamazepine ¹ , phenytoin ¹ phenobarbitone
absence	valproate	lamotrigine, clonazepam
atypical absence	valproate	lamotrigine
atonic, clonic		topiramate, clonazepam
myoclonic	valproate, clonazepam	topiramate, phenobarbitone piracetam, levetiracetam lamotrigine ²
infantile spasms	adrenocorticotrophin (ACTH), prednisolone, vigabatrin ³	nitrazepam, clonazepam valproate

footnote: 1. may aggravate myoclonus/absence seizure in Idiopathic Generalised Epilepsy

2. may cause seizure aggravation in SMEI and JME

3. especially for patients with Tuberosc sclerosis

Table 5. Side effects and serious toxicities of anticonvulsants.

seizure type	common side effects	serious toxicity
carbamazepine	drowsiness, dizziness, ataxia, diplopia, rash	agranulocytosis Steven-Johnson syndrome ¹
clonazepam	drowsiness, hypotonia, salivary and bronchial hypersecretion, paradoxical hyperactivity and aggressiveness	
lamotrigine	dizziness, somnolence, insomnia, rash	Steven-Johnson syndrome
levetiracetam	somnolence, asthenia, dizziness, irritability, behavioural change	
phenobarbitone	behavioural disturbance, cognitive dysfunction, drowsiness, ataxia, rash	
phenytoin	ataxia, diplopia, dizziness, rash, sedation hirsutism, gum hypertrophy	megaloblastic anemia
topiramate	weight loss, somnolence, mental slowing, word-finding difficulty, hypohidrosis	renal calculi
valproate	nausea, epigastric pain, tremor, alopecia, weight gain, hair loss, thrombocytopaenia	hepatic toxicity (< 2 years age) hepatitis, pancreatitis, encephalopathy
vigabatrin	drowsiness, dizziness, mood changes, weight gain	peripheral visual field constriction (tunnel vision)

footnote: 1. Steven-Johnson syndrome occur more frequently in children of Chinese and Malay ethnicity

The patients with “Intractable Epilepsy”

Please re-evaluate for the following possibilities:-

- as it a seizure or non-epileptic event?
- anticonvulsant dose not optimised
- poor compliance to anticonvulsant
- wrong classification of epilepsy syndrome, thus wrong choice of anticonvulsant
- anticonvulsant aggravating seizures
- lesional epilepsy, hence a potential epilepsy surgery candidate
- progressive epilepsy or neurodegenerative disorder

When to refer to a Paediatric Neurologist?

- poor seizure control despite monotherapy with 2 two different anticonvulsants
- difficult to control seizures beginning in the first year of life
- seizures and developmental regression
- structural lesion on neuroimaging

Advice for Parents

- educate and counsel on epilepsy
- emphasize compliance if on anticonvulsant.
- don't stop the medication by themselves. This may precipitate breakthrough seizures.
- in photosensitive seizures - watch TV in brightly lit room. Avoid sleep deprivation.
- use a shower with bathroom door unlocked.
- no cycling in traffic, climbing sports or swimming alone.
- know emergency treatment for seizure.
- inform teachers and school about the condition

Table 6. Anticonvulsants that aggravate seizure types

phenobarbitone	absence seizures
clonazepam	tonic status in LGS
carbamazepine	absence, myoclonic, GTC seizures
lamotrigine	SMEI myoclonic seizures in JME
phenytoin	absence, myoclonic seizures
vigabatrin	myoclonic, absence seizures
<i>GTC, generalised tonic-clonic; LGS, Lennox-Gastaut syndrome; SMEI, severe myoclonic epilepsy of infancy; JME, juvenile myoclonic epilepsy</i>	