

Non-pharmacological management

Ketogenic diet *(continued)*

- ✓ A ketogenic diet should be tried for at least 3 months to assess efficacy, with consideration of continuation based on risk and benefits at each visit and after 2 years of continuous use.

In children with glucose transporter 1 deficiency syndrome or pyruvate dehydrogenase complex deficiency the diet should be continued into adulthood (lifelong treatment).

Surgery

- R Children with drug-resistant epilepsy who fulfil referral criteria for assessment for surgery should be identified early.

Vagus nerve stimulation

- R Vagus nerve stimulation could be considered as an adjunctive treatment for children with drug-resistant epilepsy who are not candidates for surgery, under the specialist guidance of a consultant paediatric neurologist.

Cognitive, developmental and psychiatric comorbidities

- ✓ Where there is evidence of severe and persistent impairments in cognitive functioning or where difficulties are less well understood, healthcare professionals should refer for specialist neuropsychological assessment.

- R Healthcare professionals should routinely enquire about depression and anxiety symptoms in all children and young people with epilepsy.

- R Cognitive behavioural therapy focusing on depression could be considered in children and adolescents with epilepsy and comorbid depression.

- R Treatment with selective serotonin reuptake inhibitors could be considered in children and adolescents with epilepsy and comorbid depression.

Transition

- R Paediatric services providing care to children and young people should consider the use of a planned, structured, educational approach directed at both patients and carers, to help prepare young people with epilepsy for the move to adult healthcare services.

Mortality

- R At or around the time of diagnosis healthcare professionals caring for children and young people with epilepsy should:
 - have a face-to-face discussion about SUDEP with families/carers and young people
 - provide written information to reinforce information provided face-to-face.

Sources of useful information

Epilepsy Scotland

Helpline: 0808 800 2200

www.epilepsyscotland.org.uk

Email: enquiries@epilepsyscotland.org.uk

Epilepsy Connections

Tel: 0141 248 4125

www.epilepsyconnections.org.uk

Email: epilepsyconnections.org.uk

Matthew's Friends

Tel: 01342 836571

www.matthewsfriends.org

Email: enq@matthewsfriends.org

SUDEP Action

www.sudep.org

Email: info@sudep.org

Young Epilepsy

www.youngepilepsy.org.uk

Email: info@youngepilepsy.org.uk

SIGN 159

Epilepsies in children and young people: Investigative procedures and management

Quick reference guide

May 2021

This Quick Reference Guide provides a summary of the main recommendations in **SIGN 159 Epilepsies in children and young people: Investigative procedures and management**.

Recommendations **R** are worded to indicate the strength of the supporting evidence. Good practice points ✓ are provided where the guideline development group wishes to highlight specific aspects of accepted clinical practice.

Details of the evidence supporting these recommendations can be found in the full guideline, available on the SIGN website: www.sign.ac.uk.

This guideline provides recommendations based on current evidence for best practice in the investigative procedures and management of epilepsies in children and young people.

Investigative procedures

- ✓ In any child being evaluated for paroxysmal events of uncertain nature a typical event should be captured on video, when safe to do so, for review by a clinician with expertise in epilepsy.

Electroencephalogram

- R** If a clinical diagnosis of epilepsy has been made, EEG is recommended for further classification of epilepsy. If standard EEG is normal, a second-line EEG that captures sleep should be carried out. This could be an ambulatory, sleep-deprived or melatonin-induced sleep EEG.

Magnetic resonance imaging

- R** In children with drug-resistant focal epilepsy, 3-T MRI should be considered if 1.5-T MRI does not detect and define a lesion.

Genetic testing

- ✓ Genetic testing should be considered, discussed and offered to the families of all children and young people presenting with epilepsy for whom aetiology cannot be fully explained through history taking, examination, targeted metabolic tests or neuroimaging. Families should be counselled by an experienced professional before genetic testing is undertaken.

Non-pharmacological management

Ketogenic diet

- R** A ketogenic diet should be offered as a treatment option in children with drug-resistant epilepsy.
- R** A ketogenic diet is recommended in children with glucose transporter 1 deficiency syndrome and should be started as soon as possible after diagnosis.
- R** A ketogenic diet should be considered for children:
- who have not responded to two antiepileptic drugs
 - with pyruvate dehydrogenase complex deficiency, ideally as part of a clinical trial with monitoring
 - with drug-resistant myoclonic-atonic epilepsy
 - and infants with infantile spasms who have not responded to standard treatment
 - with drug-resistant Dravet syndrome.

(Continued overleaf)

Recommendations for pharmacological management

Seizure type	First-line therapy	Adjunctive therapy
Focal	Carbamazepine or lamotrigine could be considered. Levetiracetam, oxcarbazepine or sodium valproate could be considered if carbamazepine and lamotrigine are not suitable or tolerated.	Carbamazepine, clobazam, gabapentin, lamotrigine, levetiracetam, oxcarbazepine, sodium valproate, topiramate or zonisamide (over 6 years of age) can be considered as adjunctive therapies if first-line therapies are ineffective or not tolerated. Perampanel could be considered as adjunctive therapy in adolescents from 12 years of age.
Absence	Ethosuximide should be considered as first-line monotherapy for the treatment of patients with childhood absence epilepsy. Sodium valproate should also be considered, but has a higher risk of adverse events. Lamotrigine could be considered for patients with childhood absence epilepsy if ethosuximide and sodium valproate are ineffective, not suitable or not tolerated. A combination of two or three AEDs could be considered if two first-line AEDs are ineffective. If treatment is still ineffective, advice should be sought from, or the patient should be referred to, a tertiary epilepsy specialist, to consider the use of clobazam, clonazepam, levetiracetam, topiramate or zonisamide.	
Lennox–Gastaut syndrome	Sodium valproate could be considered.	Rufinamide (4 years and older), clobazam (2 years and older), lamotrigine (2 years and older) or topiramate (2 years and older) could be considered as adjunctive therapy. Cannabidiol could be considered as an adjunctive therapy in conjunction with clobazam for children 2 years and older.
Infantile spasms/West syndrome	Hormonal treatment (adrenocorticotrophic hormone, tetracosactide or prednisolone) or vigabatrin could be considered. Children should be closely monitored for adverse events.	
Tuberous sclerosis		Everolimus could be considered for children (age 2 years and older) with refractory seizures associated with tuberous sclerosis complex, when other treatments have failed. Children prescribed everolimus should be closely monitored for adverse events.
Infantile spasms with tuberous sclerosis	Vigabatrin should be considered. Children prescribed vigabatrin should be closely monitored for adverse events.	
Dravet syndrome	Sodium valproate or topiramate could be considered.	Stiripentol or clobazam could be considered as an adjunctive therapy for children (3 years and older) with Dravet syndrome whose seizures are poorly controlled with sodium valproate. Cannabidiol could be considered as an adjunctive therapy in conjunction with clobazam for children (2 years and older).

Note: Sodium valproate should not be used in girls of childbearing potential unless there is no suitable alternative and a pregnancy prevention programme is in place.