CHAPTER 13

THE NERVOUS SYSTEM

13.1 EPILEPTIC SEIZURES

G40.6-7; G41.0-2; G41.8-9; R56.8

DESCRIPTION

An epileptic seizure is a change in movement, attention or level of awareness that is sustained or repetitive and occurs because of abnormal and excessive neuronal discharge within the brain.

LoE:IVb[1]

Epileptic seizures should be differentiated from:

» Collapse, e.g., syncope, anoxic seizures, transient ischaemic attack, cardiac arrhythmias;
» Movement disorders, e.g., paroxysmal dyskinesias, tic disorders;
» Mental health conditions, e.g., functional/dissociative seizures (also called psychogenic non-epileptic seizures), rage reactions, panic attacks, daydreaming or inattention;
» Sleep-related conditions, e.g., parasomnias, narcolepsy;
» Migraine associated disorders, e.g., migraine with visual aura.

See https://www.epilepsydiagnosis.org/epilepsy-imitators.html for a full list of conditions which may look like an epileptic seizure.

LoE:IVb[2]

Not all persons who have an epileptic seizure have epilepsy. Specific criteria must be met to diagnose epilepsy (See Section 13.4: Epilepsy).

DIAGNOSIS

Epileptic seizures are diagnosed clinically, through eye-witness accounts, videos, careful observation by the healthcare professional, and a history from the patient of the symptoms, signs and behaviours experienced prior to, and during the seizure. Epileptic seizures are classified by the International League Against Epilepsy (ILAE) into three types: focal, generalised, and unknown (see Figure 1 below). The evolution of the seizure (how it starts and progresses clinically) directs special investigations to determine the cause of the seizure and related management.

LoE:IVb[3]

Seizure types

Focal seizures

The epileptic activity arises from a particular focus, or networks limited to one hemisphere of the brain.

Focal seizures may present with motor signs (e.g., rhythmic jerking of one limb, automatisms such as lip-smacking) or with non-motor signs (e.g.,

olfactory, tactile, or visual hallucinations, or intense emotions such as fear). This depends on the site of origin, which may be the frontal lobe, temporal lobe, parietal lobe or occipital lobe. A focal brain lesion should always be excluded in new focal seizures.

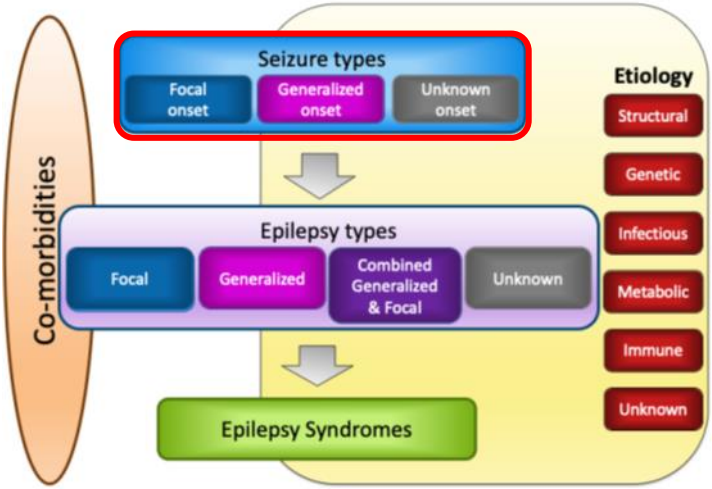


Figure 1. International League Against Epilepsy classification of seizure types *(Source: Scheffer IE, Berkovic S, Capovilla G, Connolly MB, French J, Guilhoto L. ILAE classification of epilepsies: Position paper of the ILAE Commission for Classification and Terminology. Epilepsia. 2017, 58 (4): 512-521)*

LoE:IVb[4]

Focal seizures are classified according to the degree of impaired consciousness and whether there is progression to a tonic-clonic seizure. Consciousness is evaluated by assessing the levels of awareness (of themselves and their surroundings) and responsiveness (to other people or stimuli) of the person during the seizure. Any impairment in consciousness means that the person's safety and the safety of others must be protected during the seizure.

» **Focal preserved consciousness seizures:** (previously termed 'simple partial seizures'): the person is fully aware of themselves and their surroundings and fully responsive to others throughout the seizure.
» **Focal impaired consciousness seizures:** (previously termed 'complex partial seizures'), the person has impaired awareness or responsiveness at any time during the seizure.
» **Focal unknown state of consciousness seizures:** used when the state of consciousness is not known (e.g. unclear information).
» **Focal-to-bilateral tonic-clonic seizure:** the epileptic seizure progresses to both brain hemispheres. Bilateral tonic-clonic seizures are differentiated from generalised tonic-clonic seizures by a history of preceding focal signs (either sensory or motor) occurring before complete loss of consciousness

and the development of tonic-clonic movements. The terms 'aura' or 'warning signs' may be used by people for the focal signs of the seizure.

Generalised seizures
The epileptic activity arises within and rapidly spreads to involve networks in both hemispheres of the brain.
Generalised seizures are almost always associated with impaired or loss of consciousness. Generalised seizures are classified as:

» **Generalised motor seizures**, which include:
  – **Generalised tonic-clonic seizures**, with loss of consciousness and bilateral tonic-clonic limb movements.
  – **Generalised seizures other than tonic-clonic**, including seizures with varying degrees of impaired consciousness and bilateral *tonic* movements (stiffening, sometimes with vibratory movements) of limbs or eyes; bilateral *atonic* movements (sudden loss of muscle tone) of head, trunk or limbs; or bilateral jerks (brief shock-like muscle contractions), as in *myoclonic* seizures.

» **Absence seizures** (previously termed 'petit-mal seizures'), which usually occur in association with an epilepsy syndrome (See Section 13.4: Epilepsy). Absence seizures may be:
  – **'typical'** with abrupt loss of consciousness lasting 5-30 seconds and clonic movements of face and/or automatisms, or
  – **'atypical'** with a less abrupt onset of impaired consciousness, longer seizure duration, and loss of muscle tone of head, trunk and limbs. Atypical absence seizures are rare and can be challenging to differentiate from focal sensory seizures.

LoE:IVb[5]

Unknown:
The category of 'unknown onset' is used when there is not enough information, or the clinical presentation is too unclear, to distinguish between focal or generalised seizures.

For more detail and educational videos on seizure types, see https://www.epilepsydiagnosis.org/seizure/seizure-classification-groupoverview.html

13.2 STATUS EPILEPTICUS

G41.2; G41.8-9

DESCRIPTION

In status epilepticus, the seizures do not stop, or they occur repeatedly in close succession with impaired consciousness between seizures. Status epilepticus may be 'convulsive' (associated with prominent motor symptoms) or 'non-convulsive' (i.e., without prominent motor symptoms).

Convulsive status epilepticus:

Convulsive status epilepticus is defined as ≥ 5 minutes of either:

» continuous generalised, or bilateral tonic clonic seizure, or
» two or more discrete generalised or bilateral tonic clonic seizures with incomplete recovery of consciousness between the seizures.

Convulsive status epilepticus is a **medical emergency**. There are two critical time points:

» Time point 1: **5 minutes** from the onset of the initial epileptic seizure (i.e., at the point of diagnosis). Immediate treatment is needed to prevent ongoing epileptic seizure activity.
» Time point 2: **30 minutes** of epileptic seizure activity, timed from the onset of the seizure. After 30 minutes of seizure activity, irreversible brain damage related to hypoxia, acidosis, depletion of local energy stores, cerebral oedema and structural damage, is likely to occur.

Complications of convulsive status epilepticus include:

» hyperpyrexia
» respiratory depression
» cerebral oedema
» blood pressure disturbances
» inappropriate antidiuretic hormone (ADH) secretion
» hypoxic ischaemic damage to brain, myocardium and muscles.
» disturbances of blood glucose
» renal failure
» acidosis

Non-convulsive status epilepticus:

Non-convulsive status epilepticus refers to abnormally prolonged or rapidly recurring epileptic seizures with impaired consciousness but no major motor symptoms (e.g., focal seizures with autonomic, sensory or perceptual manifestations or absence seizures). The presentation is often subtle, and the seizures may not be recognized.

Diagnosis is confirmed on EEG.

Treat as for convulsive status epilepticus below (See Section 13.2.1. Epileptic seizures and status epilepticus in children < 13 years of age, and Section 13.2.2. Epileptic Seizures and Status Epilepticus in Adolescents (13 – 18 Years). Ensure that underlying causes are identified and managed.

CAUSES OF EPILEPTIC SEIZURES AND STATUS EPILEPTICUS

With every epileptic seizure, the underlying cause of the seizure must be determined and treated, including in people with epilepsy.

Important causes of epileptic seizures that must be considered include:

» Infectious conditions, e.g., meningitis or encephalitis.
» Encephalopathy, e.g., hypertensive encephalopathy or cerebral hypoxia
» Metabolic conditions e.g., hypoglycaemia, hypo- or hypernatraemia, hypocalcaemia.
» Brain lesions, e.g., brain tumours, stroke and post-stroke sequelae, trauma and post-traumatic sequelae.

» Substance withdrawal, e.g., alcohol or benzodiazepines.
» Substance intoxication, e.g., cocaine or amphetamines.
» Poisoning or toxin ingestion (accidental or intentional as in an overdose), e.g., isoniazid.
» Other neurological (e.g., cerebral palsy) or neurodegenerative (e.g., Alzheimer's dementia) conditions.
» Epilepsies, e.g., breakthrough seizures, treatment non-adherence, recent changes to antiseizure medicine (ASM), antiseizure medication toxicity.

13.2.1 EPILEPTIC SEIZURES AND STATUS EPILEPTICUS IN CHILDREN < 13 YEARS OF AGE

DESCRIPTION

Central nervous system infections are a common cause of epileptic seizures in children under 13 years of age in South Africa. The most common seizures in children are febrile seizures (See Section 13.3: Febrile seizures) but history, examination and investigations must aim at excluding the following conditions:

Perinatal conditions	Infections	Poisoning
» congenital infection » hypoxic-ischaemic damage » trauma » cerebral haemorrhage or thrombosis	» meningitis » encephalitis » brain abscess » neurocysticercosis	» medicine toxicity, e.g., isoniazid, antihistamines, antiseizure medicines » substance abuse, e.g., solvents, amphetamines » environmental and other toxins e.g., pesticides (organophosphates), essential oils » substance withdrawal, e.g. benzodiazepines
Metabolic abnormalities	**Systemic disorders**	**Primary cerebral causes**
» hypoglycaemia » hypocalcaemia » hypomagnesaemia » hyponatraemia » hypernatraemia » inborn errors of metabolism	» vasculitis » hypertensive encephalopathy » uraemia (renal failure) » hyperammonaemia (liver failure)	» cerebral malformation » genetic/familial (syndromic) » tumour » idiopathic/ unknown

Special considerations by age group

Neonates:

Neonatal seizures are usually provoked, most commonly by hypoxic ischaemic encephalopathy (HIE). They rarely lead to a subsequent diagnosis of epilepsy. For management, refer to Section 19.6.2 Seizures, Neonatal.

Infants and children up to 2 years of age:

» The most frequent cause of epileptic seizures that present in infancy are febrile seizures (See Section 13.3: Febrile seizures).
» Infants under 6 months of age cannot manifest generalised tonic-clonic seizures.
» Most provoked seizures presenting in this age group appear to be focal, even when a generalised brain pathology is present.
» Infants and children under 2 years may not have typical signs of meningitis, i.e. neck stiffness or Kernig sign. Meningitis should be excluded in all children with a fever and seizure.
» Infants and children are at risk of metabolic derangement which may be from common triggers (e.g., dehydration) or rare causes (e.g., inborn errors of metabolism).
» Neuroimaging is often useful, especially to exclude structural aetiologies (e.g., intracranial bleeds).

Children 2 to 12 years of age:

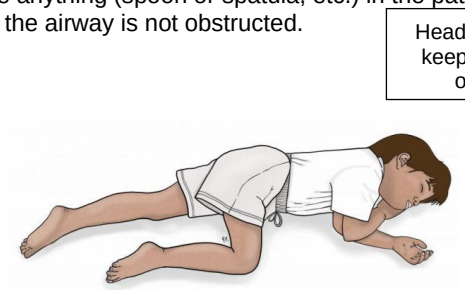
Older children are often able to present with clearer descriptions of seizure onset, i.e. a focal onset seizure is more likely to be specifically driven by an underlying focal cause (potentially a structural pathology):

» Ask about focality – point to the side of movement or change.
» Change in behaviour before convulsive seizure becomes evident.
» Altered consciousness.
» Type of seizure is related to the underlying aetiology.

GENERAL MEASURES

On arrival/ while fitting:

» Ensure the environment is safe. Remove all sharp objects and hot liquids.
» Place the child in a recovery position to prevent aspiration of secretions or vomitus, on the floor if necessary.
» Do not place anything (spoon or spatula, etc.) in the patient's mouth.
» Ensure that the airway is not obstructed.



Adapted from https://www.kidshealth.org.nz/emergencies/emergencies-cpr

LoE:IVb[6]

» Administer oxygen via face mask or nasal cannula to maintain $SaO_2 \geq 95\%$.
» Obtain eyewitness account of when the seizure started and associated impaired consciousness: **If seizure duration is ≥ 5 minutes, commence urgent medicine treatment for convulsive status epilepticus** (refer to table below on medicine management and supportive care of status epilepticus in children < 13 years of age).
» Examine for fever; dehydration; meningism; hypoglycaemia; evidence of toxin or poison ingestion; head, neck or other trauma; obvious focal neurology; and other possible causes of the seizure.
» Establish vascular (IV or IO) access, if possible. See Section 1.1.10 Intra-Osseous Infusion in Emergencies.
» Once threat of falling is no longer present, move to bed (with cot sides are up), if needed.
» Monitor vital signs every 15 minutes.
» Keep nil per mouth.
» Ensure the family/caregiver/escort is attended to. Social worker or auxiliary staff member should obtain all contact details and provide counselling as needed.

MEDICINE TREATMENT

The aim is to control the seizure within 30 minutes of its onset, prevent complications of status epilepticus with supportive care, and identify and correct underlying causes. Follow standard resuscitation protocols, such as the ABCDE approach.

Table 1: MEDICINE MANAGEMENT AND SUPPORTIVE CARE OF STATUS EPILEPTICUS IN CHILDREN < 13 YEARS

PHASE	MEDICINE MANAGEMENT	SUPPORTIVE CARE
EARLY STATUS EPILEPTICUS **5-10 minutes after seizure onset**	**LEVEL 1 INTERVENTION:** (benzodiazepines, up to 2 doses) <u>If vascular access is available:</u> • Lorazepam, IV or IO, 0.1 mg/kg over 60 seconds (max 4 mg/dose). **OR** • Midazolam, IV or IO, 0.25 mg/kg over 60 seconds, (max 10 mg/dose). **OR** • Diazepam, IV or IO, 0.25 mg/kg IV over 60 seconds (max 10 mg/dose). *LoE:IVb*[7] <u>If vascular access is not available:</u> • Lorazepam, IM or buccal, 0.1 mg/kg (max 4 mg/dose). **OR** • Midazolam, IM, 0.1 mg/kg, or buccal*, 0.5 mg/kg. *LoE:IIIa*[8] **OR** • Diazepam, rectal**, 0.5 mg/kg (max 10 mg/dose). *LoE:IIIa*[9] Expect a response within 1–5 minutes. **If the seizure does not resolve within 5 minutes after first dose, give a repeat dose of benzodiazepine.** **CAUTION** Benzodiazepines can cause respiratory depression. Monitor oxygen saturation and respiratory rate. If respiratory depression occurs, assist ventilation with bag-valve mask (1 breath every 3–5 seconds) and refer urgently/transfer to a high care setting.	» Aim for seizure control within 30 minutes of onset. » Provide supplemental oxygen, maintain $SaO_2 \geq 95\%$. » Monitor cerebral perfusion pressure (CPP), heart rate, oxygen saturation. » Check glucose. If low, correct and start maintenance IV fluid with dextrose 5% in sodium chloride 0.9%. Do not overhydrate. » Blood gas analysis for electrolytes. Correct as required. <u>Other biochemical disorders:</u> Correct abnormalities, if present, e.g. glucose, calcium and sodium. Take blood for electrolytes, LFTs, FBC. If patient is a known epileptic, check therapeutic levels of antiseizure medications (ASM). If meningitis cannot be excluded, give: • Ceftriaxone, IM or IV, 100mg/kg/dose stat

PHASE	MEDICINE MANAGEMENT	SUPPORTIVE CARE
ESTABLISHED STATUS **10-30 minutes after seizure onset**	**LEVEL 2 INTERVENTION:** If vascular access is not available: • Phenobarphital, IM 20 mg/kg. ○ Slow IM injection **OR** if no IM formulation available: • Levetiracetam oral, crushed and given by nasogastric tube, 60 mg/kg as a single dose (Maximum dose: 4500 mg). *LoE:IIIb*[10] **OR** • Phenobarbital, oral, crushed and given by nasogastric tube, 20 mg/kg as a single dose. Note: Avoid repeating oral phenobarbital as it may take over an hour to achieve therapeutic concentrations, and repeat doses increase the risk of respiratory depression. If no response to phenobarbital IM or oral after 5-20 minutes, levetiracetam oral may be given via nasogastric tube. If vascular access is available: • Phenytoin, IV or IO, 20 mg/kg *(diluted in sodium chloride 0.9% and infused over 20 minutes, not exceeding 3 mg/kg/min with cardiac monitoring)*. **OR** • Phenobarbital, IV or IO, 20 mg/kg over 5 mins (max 600 mg/dose). **CAUTION** Do not use phenytoin to manage drug- or toxin-induced seizures. Cardiac monitoring is mandatory to ensure safe use. *LoE:IVb*[11] If phenytoin given and no response after 15–20 mins of infusion: • Phenobarbital, IV or IO, 20 mg/kg over 5 mins (max 600 mg per dose).	» Consult with higher level care and refer urgently. » Prepare for intubation and ventilation. » Ensure that phenytoin is administered independently of other IV fluid, i.e. use a separate IV line, or stop maintenance fluids, flush the line with saline and commence the phenytoin infusion. » Seizures due to poisoning should **NOT** be treated with phenytoin.

PHASE	MEDICINE MANAGEMENT	SUPPORTIVE CARE
	<u>If phenobarbital given and no response 5 mins after administration:</u> • Phenytoin, IV or IO, 20 mg/kg *(diluted in sodium chloride 0.9% and infused over 20 minutes, not exceeding 3 mg/kg/min with cardiac monitoring)*. **OR** Repeat phenobarbital, IV in half doses: • Phenobarbital, IV or IO, 10 mg/kg over 5 mins (max 600 mg/dose). <u>If still no response:</u> • Phenobarbital, IV or IO, 10 mg/kg over 5 mins (max 600 mg/dose). *LoE:IVb*[12] Prepare for intubation if sufficiently skilled in the procedure and relevant rescue devices are available. **Refer to ICU**	
REFRACTORY STATUS Seizures persist despite treatment with adequate doses of two or three antiseizure medications **USUALLY by 30- 60 minutes after seizure onset**	**Failure of level 1 and level 2 interventions to control seizures** **Refer to ICU** Consider: » Midazolam infusion. » Maintain SaO2 ≥ 95%: Oxygen, by facemask or nasal cannulae while convulsing. » Endotracheal intubation with neuroprotective ventilation strategy (See Section 23.1: Rapid sequence intubation). » If it is necessary to ventilate, maintain PaCO2 in the low-normal range, i.e. 4.0–4.5 kPa. » Measure antiseizure medication blood concentrations if there are breakthrough seizures on medication, signs of toxicity, drug interactions or concerns about adherence.	» Stop seizure. » Support haemodynamic status. » Admit to high- or intensive-care, if possible. » Monitor: > heart rate, respiratory rate, > blood pressure, SaO2; > blood gas analysis, acid-base status; > electrolytes, blood glucose, neurological status; > fluid balance, osmolality; > Blood levels of ASM. » Cardiovascular and/or respiratory support if the patient is unable to maintain blood gases and blood

PHASE	MEDICINE MANAGEMENT	SUPPORTIVE CARE
		pressure within the normal physiological range. **Cerebral oedema** Treat when clinically proven. See Section 13.12: Raised intracranial pressure.

Note:

» Watch for complications of the prolonged seizure.
» Check all possible underlining conditions.
» Watch for adverse effects of administered ASM.

* Midazolam, buccal, 0.5 mg/kg/dose. See Primary Health Care STGs and EML, Chapter 23: Standard paediatric dosing tables.

- Use midazolam for injection 5 mg in 1 mL undiluted.
- Draw up the required volume in a 5 mL syringe.
- Remove needle then administer midazolam into the buccal cavity (between gum and cheeks).
- Note: Buccal midazolam should not be used in infants < 6 months of age.

LoE:IIIa[13]

**Diazepam, rectal, 0.5 mg/kg/dose as a single dose. See Primary Health Care STGs and EML, Chapter 23: Standard paediatric dosing tables.

- Use diazepam for injection 10 mg in 2 mL undiluted.
- Draw up the required volume in a 2 mL syringe.
- Remove needle then connect syringe to an NGT and gently insert into the rectum (or insert the whole barrel of the lubricated syringe if no NGT available) and inject the contents.
- Remove NGT / syringe and hold buttocks together to minimise leakage.

AFTER THE SEIZURE:

Post-ictal phase

» Keep nil per mouth and haemodynamically stable until child has regained consciousness.
» Determine the seizure type (focal or generalised) and cause of the seizure. Further investigations are driven by clinical signs and seizure onset, e.g.:
  - History of toxin exposure
  - Evidence of neuro-infection
  - History of trauma
  - Focal onset (e.g., a warning experience prior to generalised tonic-clonic movements) and/or focal neurology warrants neuroimaging.
» If meningitis cannot be excluded, commence antibiotic therapy within one hour of arrival. Refer to Paediatric Hospital Level STGs and EML, Chapter Sections 8.11: Meningitis, acute bacterial; 8.13 Meningo-Encephalitis/Encephalitis, Acute viral. Also refer to Primary Health Care STGs and EML, Chapter 15, Section 15.8.1: Acute meningitis.

Pre-discharge

» Consider whether the criteria for a diagnosis of epilepsy are met (see Section 13.4: Epilepsy). Initiate appropriate ASM for type of epilepsy and develop an emergency care plan for recurrent seizures.
» If not epilepsy, start weaning any ASMs.
» Counsel the caregiver on the current state of the child, the reason for the seizure, management given and likely sequelae of the seizure. Offer only as much information as the caregiver can receive at that time.
» Set up a follow up appointment to re-evaluate the diagnosis, educational and social needs, and to reinforce educational counselling about the child's condition.

Active follow-up

» Ensure underlying medical conditions are appropriately managed.
» Reduce and stop any residual ASMs if not epilepsy.
» Assess neurodevelopmental conditions, educational and social needs. Refer as necessary.
» If epilepsy is the cause of the seizure, check seizure control and if emergency care plan is understood.

REFERRAL

» Refractory status/lack of High Care/ICU beds.

13.2.2. EPILEPTIC SEIZURES AND STATUS EPILEPTICUS IN ADOLESCENTS (13 – 18 YEARS)

See Adult Hospital Level STGs and EML, Chapter 14: Central Nervous System, Section 14.5.1 Epileptic Seizures and Status Epilepticus in Adolescents (13 – 18 Years) And Adults.

13.3 FEBRILE SEIZURES

R56.0

DESCRIPTION

Febrile seizures occur between the ages of 6 months and 6 years of age in association with a significant fever (>38.5°C) in the absence of an intracranial infection and are typically associated with viral upper respiratory tract infections. These are the most common type of seizures in children of this age, however, diagnosis requires the exclusion of other causes of seizures.

LoE:IVb[14]

Febrile convulsions can be simple or complex.
Simple febrile seizures:

» are generalised,
» occur once per illness,
» last less than 15 minutes (typically 1–2 minutes),
» are not associated with any neurological deficit,
» are self-limiting,
» consist of only one seizure during the febrile illness which needs no specific treatment, and
» may be associated with a family history of simple febrile seizures.

Complex febrile seizures have *one or more* of the following characteristics:

» Seizure duration of 15 minutes or more;
OR
» are recurrent within the same febrile illness;
AND/OR
» have a focal onset.
AND/OR
» are associated with post-ictal, focal neurological abnormalities.

Children with febrile seizures have a good prognosis, and very rarely develop epilepsy.

The overall risk for recurrence of simple febrile seizures is 30 - 40%. Factors which increase the risk for recurrent febrile seizures include:

» seizure disorder in a first-degree relative,
» onset before 12 months of age,
» initial complex seizures.

DIAGNOSTIC CRITERIA

Clinical

» Investigate for intracranial, extracranial, and biochemical causes of fever or seizure.
» Signs of meningism are unreliable in children less than 18 months of age.
» If raised intracranial pressure or meningitis cannot be excluded, the diagnosis of febrile seizures cannot be made. Treat children empirically for meningitis if suspected.

Investigations

Lumbar puncture

» Lumbar puncture is indicated in:
  – All children with clinical features of meningitis.
» Lumbar puncture may be indicated in:
  – Children where meningitis cannot be excluded, e.g. <18 months of age or those who have received antibiotics prior to the event.
» No further investigation is required in children ≥18 months of age where a focus of extracranial infection is present, and intracranial infection such as meningitis has been excluded clinically.

Neuroimaging

» Children with complex febrile seizures and/or persistent lethargy may require neuroimaging, followed by a lumbar puncture if raised intracranial pressure can reliably be excluded based on clinical assessment (see Section 13.11: Lumbar Puncture).
» Based on clinical findings, investigate complex febrile seizures for possible underlying conditions such as meningitis, focal brain lesions and epilepsy.

Note:
An EEG is of no value in simple febrile seizures but may be considered in recurrent complex febrile seizures.

GENERAL AND SUPPORTIVE MEASURES

» Reassure parents and caregivers.
» Educate parents and caregivers regarding the first aid management of seizures.
» Counsel against tepid sponging and fans as this increases the core temperature.
» Counsel that recurrent febrile seizures occur in 30-40% of patients.
» Look for a cause of the fever.
» **Always exclude meningitis** (See Section 8.11: Meningitis, acute bacterial).

MEDICINE TREATMENT

For fever related symptoms (temperature > 38.5°C):

- Paracetamol, oral, 15 mg/kg/dose 6 hourly.
  - Note: Paracetamol has no effect on seizure prevention.

For convulsions lasting 5 minutes or longer, see Section 13.2: Status epilepticus.

Continuous antiseizure medication prophylactic therapy.

» Routine daily antiseizure medication prophylaxis is not recommended for patients with simple febrile seizures.
» For children with recurrent complex febrile seizures, discuss the treatment options with a specialist.

REFERRAL

» All patients with recurrent complex febrile seizures without an obvious cause of the seizure and/or not responding to initial management should be discussed with a specialist.
» Developmental delay/regression.

13.4 EPILEPSY

G40.0-9

DESCRIPTION

Epilepsy is a disease of the brain defined by any of the following conditions:

» At least two unprovoked (or reflex) seizures occurring >24 hours apart, or
» One unprovoked (or reflex) seizure if there is a high risk (60% or more) of having recurrent seizures within the next 10 years (i.e., if the person is vulnerable to having another unprovoked seizure, e.g. because of structural damage such as from a stroke) or
» Diagnosis of an epilepsy syndrome.

Note:

» An “unprovoked” epileptic seizure is a seizure which does not have evidence of an identifiable temporary or reversible factor acting on a healthy brain (e.g., hypoglycaemia, alcohol withdrawal, concussion).
» A “reflex” epileptic seizure is a seizure which occurs in response to a stimulus such as flashing lights. Such epileptic seizures indicate the person’s brain is predisposed to having seizures and therefore warrant a diagnosis of epilepsy.
» Epilepsy may be diagnosed after a single unprovoked seizure in people with an increased risk of recurrence for example in people with previous conditions such as TB meningitis, neurocysticercosis, stroke, brain tumour or traumatic brain injury. Note that the single unprovoked seizure is not caused by the immediate insult to the brain but occurs spontaneously (i.e., is unprovoked) because of the long-term sequelae of the initial insult. The damaged brain is thus at high risk of a recurrent unprovoked epileptic seizure.
» Epileptic syndromes confer a diagnosis of epilepsy, even if the risk of recurrent epileptic seizures is low for a particular individual.
» Epilepsy is considered to be resolved and no longer needing maintenance treatment in individuals who either:

- had an age-dependent epilepsy syndrome, but are now past the applicable age, **OR**
- have remained seizure-free for the last 10 years and weaned off ASM for at least 5 years.

» Epilepsy is associated with many psychological, social and legal problems, as well as cultural misperceptions which should be explored and addressed at the time of diagnosis, and throughout the course of the illness.

Epilepsy types

As shown in Figure 2, epilepsies are classified by the International League Against Epilepsy (ILAE) according to:

» Type of seizures experienced, e.g., focal, generalised, combined generalised and focal, or unknown.

AND

» Aetiology, which may be:

- Structural (e.g., cerebral or vascular malformations, stroke, traumatic brain injury, brain tumours).
- Genetic (the epilepsy is a direct result of chromosomal or gene abnormalities, e.g., Down syndrome, Fragile X syndrome, Dravet syndrome).
- Infectious (e.g., post-infectious sequelae of TB meningitis).
- Metabolic (e.g., inborn errors of metabolism).
- Immune (rare conditions involving neuroreceptor antibodies).
- Unknown.

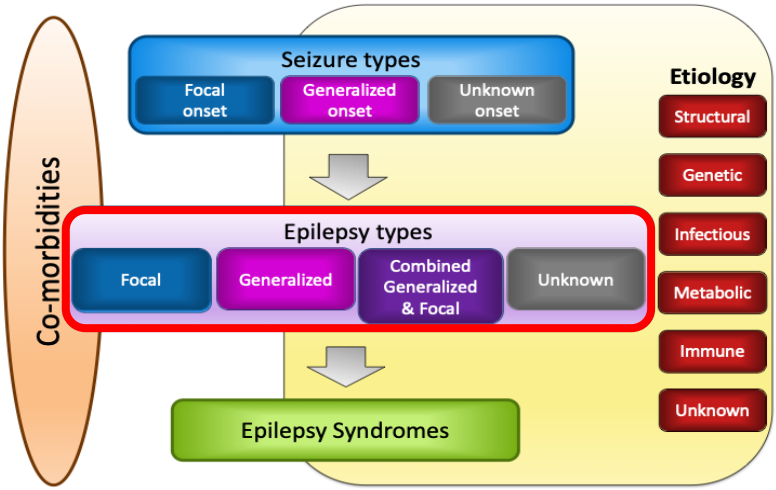


Figure 2. International League Against Epilepsy classification of seizure types *(Source: Scheffer IE, Berkovic S, Capovilla G, Connolly MB, French J, Guilhoto L. ILAE classification of epilepsies: Position paper of the ILAE Commission for Classification and Terminology. Epilepsia. 2017, 58 (4): 512-521)*

LoE:IVb[15]

Focal epilepsy
Characterised by unprovoked focal seizures, which may or may not evolve to bilateral tonic-clonic seizures. The diagnosis is made clinically and requires a detailed description of how the seizure started. In people presenting with generalised tonic-clonic seizures, it is important to ask about any warning symptoms or 'aura' experienced by the person before losing consciousness. Typical interictal and/or ictal EEG findings may be present, and neuroimaging may reveal a focal brain lesion, supporting the diagnosis, but may also be normal.

Generalised epilepsy
Characterised by unprovoked generalised seizures, including tonic-clonic, tonic, myoclonic, and absence seizures. Typical interictal and/or ictal EEG findings may be present.

Combined generalised and focal epilepsy
Diagnosed in people with more than one type of seizure, e.g. unprovoked focal seizures and unprovoked generalised seizures. This may occur in people with Dravet syndrome or Lennox-Gastaut syndrome.

Unknown epilepsy
This classification is used when it is not possible to determine whether the epilepsy is focal, generalised, or combined generalised and focal epilepsy from the available history, clinical, and investigative findings.

For seizure types, see Section 13.1: Epileptic seizures.
For more information and educational videos on epilepsy types, see https://www.epilepsydiagnosis.org/epilepsy/epilepsy-classification-groupoverview.html

Investigations:

» Neuroimaging (a CT Brain or MRI if available) should be conducted:
  – in new focal onset seizures to exclude a focal brain lesion.
  – if the epilepsy features change in an individual (i.e., new symptoms appear, noting that most people will experience the same march of symptoms with each seizure).
  – if epileptic seizures recur despite adherence to treatment and the diagnosis is unclear.
» EEG is indicated for recurrent or syndromic seizures where a diagnosis cannot be made on clinical grounds alone. Delay an EEG for at least one week after the convulsive episode.
» EEG is not indicated for simple febrile seizures.
» If the seizure presentation is atypical, a 12-lead ECG should be considered to identify prolonged QT interval syndromes. Syncope with exercise, syncope in response to loud noise, fright, or extreme emotional stress, syncope whilst supine, a family history of sudden death in a young

person, e.g., less than 40 years old, or sensorineural deafness are associated with some types of long QT syndrome.

13.4.1 EPILEPSY IN CHILDREN <13 YEARS OF AGE

A child may be diagnosed with focal, generalised, combined generalised and focal, or unknown epilepsy types.

Special considerations

Seizures in neonates

Seizures in the neonatal period seldom confer a diagnosis of epilepsy. For management of seizures, See Section 19.6.2: Seizures, Neonatal.

Underlying conditions and co-morbidities

The sequelae of neurocysticercosis and TB meningitis are a common cause of childhood epilepsy in South Africa. See Section 13.6: Neurocysticercosis and Section 10.4: Meningitis, Tuberculosis in Children.

Common co-morbidities with epilepsy which need to be managed in conjunction with treatment of the epilepsy include:

» Neurodevelopmental disorders, e.g., autism spectrum disorder, intellectual disability, attention deficit hyperactivity disorder, specific learning disorders.
» Behavioural and psychiatric disorders, e.g., anxiety disorder, depression, oppositional defiant disorder, conduct disorder, sleep disorders.
» Cognitive and academic difficulties.
» Developmental delays, particularly with epilepsies that have their onset in early childhood.

Infantile epileptic spasms

Infantile epileptic spasms (previously called infantile spasms) are a type of seizure that typically occur in infants between 3 and 12 months of age. They involve brief, repetitive stiffening of the body (for 1-2 seconds), typically recurring in clusters, and usually happen when the infant is waking up or falling asleep.

These spasms are often associated with a severe epilepsy syndrome called West syndrome or Infantile Epileptic Spasms Syndrome, which includes:

» Epileptic spasms
» Developmental delay or regression
» A specific abnormal EEG pattern called hypsarrhythmia

Infantile spasms are a **medical emergency.** Early diagnosis and treatment are crucial to prevent long-term developmental problems. Discuss with a specialist and refer immediately.

See Section 13.4.1.1: Epilepsy syndromes for additional information.

Children living with HIV on ART

Drug interactions between ASM and ARVs can arise from several mechanisms, including liver metabolism (increased or decreased) and competition for protein binding. There is a lack of strong evidence to guide clinicians at present.

The following points are important to remember when treating epilepsy in patients on ART:

» Carbamazepine, phenobarbital and phenytoin induce the hepatic Cytochrome P450 and glucuronidation hepatic enzyme systems, and may lead to sub-therapeutic ARV plasma levels, especially INSTIs, NNRTIs and PIs. If clinically indicated, monitor ASM levels in patients taking concurrent ART and ASM therapy.
» Consider lamotrigine, levetiracetam or valproate, and avoid prescribing carbamazepine, phenytoin or phenobarbital.

Girls likely to need treatment when of child-bearing potential/ after 10 years of age

» The potential for child-bearing must be considered in all girls requiring treatment for epilepsy.
» Choose the ASM with the least potential for harm in pregnancy (see Figure 3 below).
» Valproate should not be used if the child is likely to require epilepsy treatment as they grow older / develop child-bearing potential, unless other ASMs are not effective or intolerable. Annual acknowledgment of risk must be obtained if valproate is used.
» Girls over the age of 10 years should be counselled regarding sexual activity and contraception needs.
» Refer any girls who may be vulnerable to sexual abuse to a social worker.

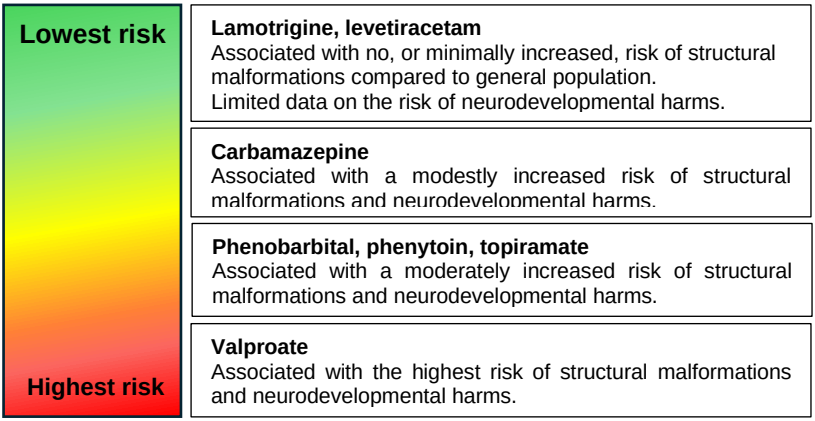


Figure 3. Risk of congenital structural malformations and neurodevelopment harms associated with various antiseizures medicines. Increasing risk refers to increasing number of pregnancies or children affected. (Adapted from Pennell PB. *Neurotherapeutics*. 2016 and Medicines & Healthcare products Regulatory Agency safety leaflet).

LoE:IVb[16]

GENERAL AND SUPPORTIVE MEASURES

» Offer ongoing counselling to the patient and caregiver about epilepsy, its associated complications and comorbidities, e.g., learning difficulties, ADHD, anxiety, and aggression.

» Encourage participation in activities of daily living and ensure that the patient is not precluded from school and lifestyle activities, unless valid safety concerns are evident.

MEDICINE TREATMENT

The goal of medicine treatment is to prevent recurrent seizures and optimise quality of life balanced against potential adverse effects of ASM. Being seizure-free may not be achievable in all children and may require individualised treatment goals, e.g., severe to profound intellectual disability with intractable seizures that do not interfere with the care of that child. Cautious escalation of treatment should be done with consideration for potential adverse effects. In the setting of apparent drug-resistant epilepsy, revisit the diagnosis, check adherence, consider if the prescribed ASM and dose are correct, and refer to specialist if treatment goals are still not achieved.

Acute therapy

Manage acute seizure and status epilepticus as per seizures/status epilepticus, see Sections 13.1: Epileptic Seizures, and 13.2: Status epilepticus.

Maintenance therapy

Principles of management

» Monotherapy is preferred.
» Combination therapy, if necessary, should be specialist initiated in the form of add-on management.
» As a general rule, start with low doses according to the lower dose per kilogram for the child and titrate upwards slowly until seizures are controlled. This is often at the low-to-mid-therapeutic dose range.
» If seizures continue, titrate to high therapeutic doses, monitoring for unacceptable adverse effects.
» If the patient experiences unacceptable adverse effects, verify that the ASM is being given correctly and that the epilepsy type has been correctly identified, then consider cross-titrating to another ASM.
» ASMs can interact with other drugs, affecting their effectiveness and safety. These interactions mainly occur through changes in drug metabolism (enzyme induction or inhibition) and protein binding.
» Monitoring of therapeutic ASM levels is only indicated when there is concern about toxicity or adherence. It is not recommended when titrating to establish the appropriate dose.

Continuation and cross-titration of treatment

Cross-titration may be required when changing from one ASM to another if there is poor control or adverse effects. Add on the new medication and up-titrate to a therapeutic dose. When seizures are controlled, slowly reduce and stop the initial medication (doses should only be changed at 2 weekly intervals).

In most children it is appropriate to continue effective ASM therapy as they grow older as indicated by the type of epilepsy. Exceptions are:

» **Valproate in girls approaching puberty and child-bearing potential**. Valproate should be deprescribed by cross-titration onto another suitable ASM in girls at approximately 10 years of age. **Lamotrigine and levetiracetam are the safest ASMs in pregnancy** and therefore the first choices in girls and women of child-bearing potential. Carbamazepine and topiramate are both teratogenic and should be avoided.
» **Phenobarbital** may be initiated in infants under 6 months of age or children with severe or profound intellectual disability, for whom the adverse effects will not interfere with their care or activities of daily living. In children who are established on phenobarbital, cross-titration onto another suitable ASM should be undertaken by 2 years of age.

Table 2: Epilepsy treatment in children 1 month to ≤ 12 years

Epilepsy type		Population	1st line	2nd line	3rd line *(specialist consultation)*	Comments
Focal epilepsy	With or without evolution to bilateral tonic-clonic seizures *LoE:IIIb*[17]	All	Lamotrigine	Carbamazepine **OR** Levetiracetam	Consider combination therapy, or add-on topiramate	Avoid carbamazepine in children with HIV on ART due to drug-drug interactions Avoid carbamazepine and topiramate in girls who may require treatment when/ if of child-bearing potential.
Generalised epilepsy	Tonic-clonic, atonic, clonic, or tonic seizures	» Boys » Girls unlikely to need treatment after age 10 years or to develop child-bearing potential	Lamotrigine (low risk*) **OR** Levetiracetam (high-risk*)	Levetiracetam or lamotrigine (whichever not used as 1st line) **OR** Valproate	Consider combination therapy with add-on Lamotrigine, **OR** Levetiracetam, **OR** Valproate, **OR** Topiramate	Valproate should not be used unless lamotrigine and levetiracetam are poorly tolerated or ineffective. Avoid topiramate in girls who are likely to require treatment when/ if of child-bearing potential. If valproate used in girls 10 years and older - see note below on **acknowledgement of risk form**.
		Girls likely to need treatment after age of 10 years	Lamotrigine (low risk*) **OR** Levetiracetam (high-risk*)	Levetiracetam **OR** lamotrigine (whichever not used as 1st line) **OR** Consider combination	Consider: Valproate **OR** Add-on Topiramate	Valproate should not be used unless lamotrigine or levetiracetam, alone or in combination, are ineffective or poorly tolerated.

Epilepsy type	Population	1st line	2nd line	3rd line *(specialist consultation)*	Comments
			therapy with lamotrigine and levetiracetam.		**If valproate is used, see note below on "Acknowledgement of risk form"** and effective family planning.
Absence	» Boys » Girls unlikely to need treatment after age 10 years or to develop child-bearing potential	Valproate	Lamotrigine	Levetiracetam **OR** Consider combination therapy	If valproate used in girls 10 years and older - see note below on **acknowledgement of risk form**.
	Girls likely to continue treatment after age of 10 years	Lamotrigine	Levetiracetam	Consider combination treatment **OR** Valproate	**If valproate is used, see note below on "Acknowledgement of risk form"** and effective family planning.
Myoclonic *Confirm diagnosis and discuss management with a specialist in all cases*	» Boys » Girls unlikely to need treatment after age 10 years or to develop child-bearing potential	Valproate	Levetiracetam	Consider Lamotrigine **OR** Topiramate, **OR** combination therapy	These seizures may be aggravated by phenytoin or carbamazepine. Lamotrigine may be effective but may also exacerbate myoclonic seizures. If valproate used in girls 10 years and older - see note below on **acknowledgement of risk form**.

Epilepsy type	Population	1st line	2nd line	3rd line *(specialist consultation)*	Comments
Myoclonic (Continued)	Girls likely to continue treatment after age of 10 years	Levetiracetam	Lamotrigine	Consider Topiramate **OR** Combination therapy **OR** Valproate	These seizures may be aggravated by phenytoin or carbamazepine. Lamotrigine may be effective but may also exacerbate myoclonic seizures. **If valproate is used, see note below on "Acknowledgement of risk form"** and effective family planning.

Combined generalised and focal epilepsy
OR
Unknown/unclassified
Discuss clinical presentation and management with a specialist in all cases

NOTE:

- Lamotrigine is the preferred first line treatment for all adult patients, including women of child-bearing potential, pregnant women, and people living with HIV.
- Lamotrigine requires slow dose up-titration and may not be suitable in people at high-risk of seizures.
- *Assess level of risk (high vs low) based on clinical judgement and discuss with a specialist if unsure. In general, high-risk patients are those with frequent seizures, a previous history of hospitalization for seizures, or previous history of status epilepticus. Low-risk patients are those with infrequent seizures, no previous hospitalization for seizures, and no history of status epilepticus.
- **If valproate is used, acknowledgment of risk must be obtained annually, even if not of child-bearing potential.** https://www.sahpra.org.za/wp-content/uploads/2025/05/GLF-CEM-PV-S01_v2-Valproate-Annual-Risk-Acknowledgement-Form-ARF.pdf
- **Girls aged 10 years and older should receive effective family planning and consider a long-acting form of contraception.**

LoE:IVb[18]

- Lamotrigine, oral
  Monotherapy:
  - Starting daily dose: 0.2 mg/kg/dose.
  - Increase slowly at 2 weekly intervals to 1–5 mg/kg/dose 12–24 hourly.
  - Rapid escalation is associated with adverse side-effect of skin rash.

Table 3: Dosing regimens when lamotrigine is used as add-on therapy:

	Week 1 and 2	Week 3 and 4	Maintenance dose
Add on therapy where regimen **does not include valproate** or other inducers/inhibitors of lamotrigine glucuronidation	0.3 mg/kg in one or two divided doses	0.6 mg/kg in one or two divided doses	0.6 mg/kg increments every 1–2 weeks to achieve a maintenance dose of 1–10 mg/kg (once a day or two divided doses) to a maximum of 200 mg/day.
Add-on therapy where **regimen includes ASMs that induce lamotrigine glucuronidation** *(e.g. phenytoin, carbamazepine, phenobarbital, etc.)*	0.6 mg/kg in two divided doses	1.2 mg/kg in two divided doses	1.2 mg/kg increments every 1–2 weeks to achieve a maintenance dose of 5–15 mg/kg (once a day or two divided doses) to a maximum of 400 mg/day.
Add-on therapy where **regimen includes valproate** *(regardless of other concomitant medication)*	0.15 mg/kg daily	0.3 mg/kg daily	0.3 mg/kg increments every 1–2 weeks to achieve a maintenance dose of 1–5 mg/kg (once a day or two divided doses) to a maximum of 200 mg/day.

LoE:IVb[19]

CAUTION - LAMOTRIGINE
Lamotrigine may cause Stevens-Johnson Syndrome.

- Levetiracetam, oral,
  - Infants 1 to < 6 months:
    - Initial dose: 7 mg/kg/dose twice daily.
    - Increase dosage every 2 weeks by 7 mg/kg/dose twice daily based on response and tolerability to the recommended dose of 21 mg/kg/dose twice daily.
  - Infants ≥ 6 months and children < 4 years:
    - Initial dose: 10 mg/kg/dose twice daily.

  - Increase dosage every 2 weeks by 10 mg/kg/dose twice daily based on response and tolerability to the recommended dose of 25 mg/kg/dose twice daily.
- Children ≥ 4 years to 12 years:
  - Initial: 10 mg/kg/dose twice daily.
  - Increase dosage every 2 weeks by 10 mg/kg/dose twice daily based on response and tolerability to the recommended dose of 30 mg/kg/dose twice daily.

- Carbamazepine, oral, 5 mg/kg/day (starting dose), 8–12 hourly.
  - Increase slowly by 2 mg/kg/day at 2 weekly intervals.
  - Usual maintenance total daily dose: 10–20 mg/kg/day.
  - Maximum total daily dose: 20 mg/kg/day.
  - Dosage intervals: syrup 8 hourly, tablets 12 hourly.
  - Exacerbates myoclonic seizures and absence seizures.

- Valproate, oral, 5 mg/kg/dose (starting dose), 8–12 hourly.
  - Increase by 5 mg/kg weekly to 15–20 mg/kg/day, given 8–12 hourly over 4 weeks.
  - Usual maintenance dose: 20-30 mg/kg/day.
  - Maximum total daily dose: 40 mg/kg/day.
  - Exclude liver dysfunction prior to initiating therapy (at least ALT) in children under 2 years, or if there is clinical suspicion of liver dysfunction or metabolic disease.
  - Consider hepatotoxicity if clinical signs such as nausea, vomiting, malaise, jaundice or abdominal pain develop.

- Phenobarbital, oral, 3–5 mg/kg/dose as single dose at night.
  - May be used in children under six months of age.
  - Not recommended as maintenance therapy for children older than 2 years due to undesirable side-effects such as sedation, behaviour disturbances, hyperkinesia and dependence, except in situations where there is poor adherence to other drugs.
  - Exacerbates absence seizures.

REFERRAL

» Seizures suggesting complex or syndromic epilepsy.
» All myoclonic seizures and infantile epileptic spasms at presentation.
» Mixed seizure types in a single patient.
» Focal seizures for neuroimaging if not available at the facility (MRI preferred).
» Neuroregression (loss of previously acquired milestones or skills).
» Seizures persisting despite two appropriate ASMs (drug-resistant epilepsy).

Information that should accompany each referral case.

» Number and frequency of seizures per month (or year).
» Date and time of most recent seizures.
» Detailed description of the seizures, including:

  - aura or warning sign
  - what happens during the seizure? (give a step-by-step account)
  - is the person conscious during the seizure?
  - how long do the seizures last on average?
  - what does the patient experience after the seizure e.g. weakness, loss of speech?
  - how long does this experience last?
- » Is there a family history of seizures?
- » What is the initial date of diagnosis?
- » Is there evidence of alcohol use or exposure?
- » Are there any comorbid conditions, e.g. diabetes, HIV and what medication is used?
- » What is the name and dosage of the ASM used to date?
- » Does the person return regularly for repeat of medication?

13.4.1.1 EPILEPSY SYNDROMES

G40.3-5

DESCRIPTION

An epilepsy syndrome is a specific seizure type associated with characteristic EEG and imaging features, natural history, age-dependent features, triggers, response to ASM, and prognosis. For classification of seizure types, see Figure 1 in Section 13.1: Epileptic seizures.

Pointers for the identification of an epilepsy syndrome:

- » Early-onset seizures (<1 year old)
- » Neuroregression (loss of previously acquired milestones or skills)
- » Global developmental delay
- » Intellectual disability
- » Specific seizure types
- » Drug-resistant epilepsy
- » Associated neurological symptoms, e.g. facial or limb weakness, ataxia, movement disorders
- » Strong family history of epilepsy
- » Associated neurocutaneous lesions

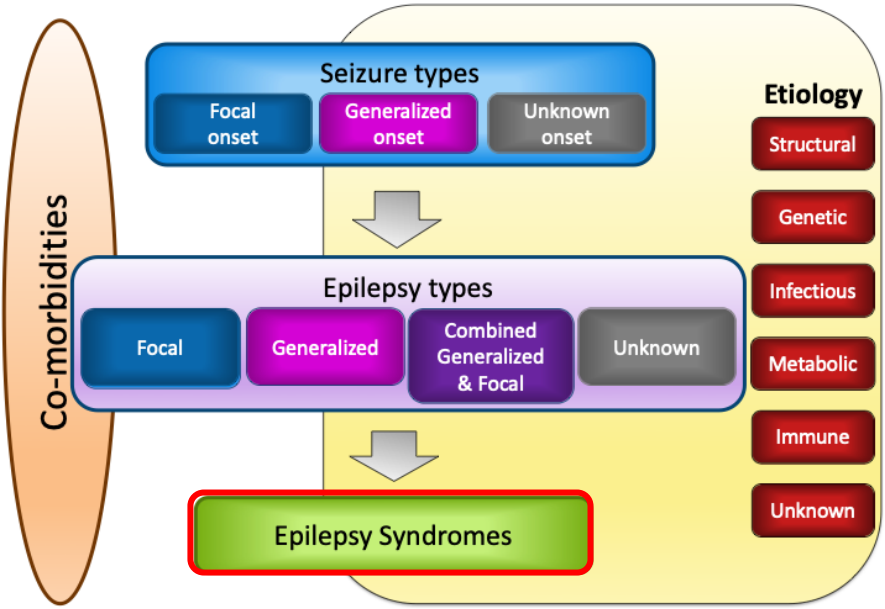


Figure 4. International League Against Epilepsy classification of seizure types *(Taken From: Scheffer IE, Berkovic S, Capovilla G, Connolly MB, French J, Guilhoto L. ILAE classification of epilepsies: Position paper of the ILAE Commission for Classification and Terminology. Epilepsia. 2017, 58 (4): 512-521.)*

Table 4: Epilepsy syndromes

Epilepsy Syndrome	Age of onset	Clinical features	Prognosis	Treatment options	Comments
Childhood Absence Epilepsy	4-10 years	Short spells of sudden onset of motor arrest and impairment of consciousness lasting between 5 and 30 seconds. Frequent episodes (up to 100/day), brief staring spells with loss of awareness, often triggered by hyperventilation.	Good; typically remits by adolescence, but some develop generalised epilepsy.	As for absence epilepsies.	**If valproate is used, see note below on "Acknowledgement of risk form" and effective family planning.**
Self-limited Epilepsy with Centrotemporal Spikes (SeLECTS)	3-13 years	Sleep-related focal seizures with hemifacial clonic spasm, e.g. facial twitching, inability to speak, hypersalivation. Awareness retained. May progress to bilateral tonic clonic seizures.	Excellent; seizures usually resolve by adolescence Refer if any deterioration in school performance Consider discontinuation of treatment if seizure free for 2 years or more, or at 14 years of age (discuss with specialist).	1st line: Lamotrigine or levetiracetam. 2nd line: Lamotrigine or levetiracetam (whichever is not used initially). 3rd line: Carbamazepine	Responds well to carbamazepine. Intermittent or rare events lacking impact on ADLs or quality of life do not require ASM unless high patient / caregiver anxiety. Carbamazepine and lamotrigine may rarely aggravate seizures.

Infantile Epileptic spasms syndrome (IESS or West syndrome)	3-6 months	Clusters of brief flexor or extensor spasms. Clinically, the child appears to stare, has sudden flexion of the trunk and head, with the limbs in extension or flexion, but held in spasm for 1-2 seconds. Events occur in clusters and are most common when the infant is going to sleep or rousing. The episodes are distressing to the infant, and he/she will often appear red in the face and may cry-out.	Poor; high risk of neurodevelopmental impairment.	Prednisone	This is a **neurological emergency**; **do not delay referral for diagnosis and treatment.** Discuss with a specialist service for support, prednisone may be started while awaiting referral on specialist advice. Early intervention reduces subsequent neuro-disability.

Lennox-Gastaut syndrome	2-3 years	Multiple seizure types (tonic, atonic, atypical absence), cognitive impairment.	Poor; seizures persist into adulthood, often drug-resistant. Behavioural problems and neuroregression occurs.	1st line: Valproate 2nd line: Lamotrigine monotherapy or add-on. 3rd line: Consider topiramate.	Tonic seizures are mandatory for diagnosis, often occur at night and should be specifically asked if present. Treat with valproate – patients are typically not of child-bearing potential. **If valproate is used, see note below on "Acknowledgement of risk form" and effective family planning.**
Dravet syndrome	Infancy, usually 3-20 months	Recurrent, prolonged febrile and afebrile seizures, later myoclonic and focal seizures.	Poor; drug-resistant epilepsy with intellectual disability and behavioural problems.	1st line: Valproate / Topiramate.	Carbamazepine, lamotrigine, and phenobarbital may aggravate seizures. Consider referral to specialist for infants under 9 months of age with unexplained, prolonged febrile or afebrile status and hemiclonic seizures.
Febrile seizures plus (FS+)	Beyond 6 years	Febrile seizures that persist beyond expected age.	Variable; some outgrow it, others develop epilepsy.	Valproate	Not all children require ASMs. **If valproate is used, see note below on "Acknowledgement of risk form" and effective family planning.**

Myoclonic atonic epilepsy (Doose syndrome)	1-5 years	Myoclonic, atonic ("drop attacks"), and generalised seizures.	Variable; small proportion of children outgrow it, others have persistent epilepsy and often association with neurobehavioural and cognitive dysfunction.	1st line: Levetiracetam 2nd line: Valproate 3rd line: Consider topiramate.	Carbamazepine and phenytoin may aggravate seizures. **If valproate is used, see note below on "Acknowledgement of risk form" and effective family planning.**

Note:

» West syndrome (IESS), Dravet syndrome, myoclonic atonic epilepsy and Lennox-Gastaut syndrome are regarded as developmental and epileptic encephalopathies (DEEs) and are associated with neuroregression/neuroplateauing, and behavioural problems.

» **If valproate is used, acknowledgment of risk must be obtained annually, even if not of child-bearing potential.** https://www.sahpra.org.za/wp-content/uploads/2025/05/GLF-CEM-PV-S01_v2-Valproate-Annual-Risk-Acknowledgement-Form-ARF.pdf

» **Girls aged 10 years and older should receive effective family planning and consider a long-acting form of contraception.**

MEDICINE TREATMENT
Discuss initiation of ASM with specialist.

REFERRAL
All suspected cases of epilepsy syndromes.

13.4.2. EPILEPSY IN ADOLESCENTS (13 – 18 YEARS)
G40.0-9

See Adult Hospital Level STGs and EML Chapter 14: Central Nervous System, Section 14.6.1 Epilepsy in adolescents and adults.

13.5 HEADACHES
R51

DESCRIPTION
Headache is the most common pain syndrome in children of all ages. Recurrent headaches are a common health problem and can be:
» primary, e.g. migraine, or
» secondary/symptomatic, e.g. raised intracranial pressure.

The actual perception of headache varies according to age and is influenced by factors such as experience, memory and cultural environment.

Extract from the International Classification of Headache Disorders (ICHD)

Migraine (without aura)
Five or more headaches lasting 1–48 hours (duration in children is often shorter, lasting a few hours only) fulfilling at least 2 of the following:
» bilateral or unilateral, frontal or parietal in location,
» pulsating in character,
» moderate or severe,
» aggravated by routine activity,
» nausea and/or vomiting plus photophobia and/or phonophobia during headache.

Migraine (with aura)
At least 2 attacks fulfilling at least 3 of the following:
» one or more reversible aura symptoms,
» at least one aura developing over > 4 minutes or 2 or more successive symptoms,
» no aura lasting > 1 hour,
» headache follows aura in less than 1 hour.

Episodic tension-type headache

At least 10 prior episodes, occurring less than 15 times per month and lasting 30 minutes to 7 days with at least 2 of the following:

» pressing or tightening quality,
» mild or moderate intensity,
» bilateral location,
» no aggravation by routine physical activity,
» no nausea, vomiting, photophobia or phonophobia.

Cluster headache

» Severe unilateral sharp headache associated with conjunctival injection and lacrimation.
» Rare in childhood.

Paroxysmal Hemicrania Continua

» Cluster headache of shorter duration.

Each of the above can occur in combination in any patient, i.e. mixed/comorbid headache.

Headaches can also be sub-classified according to temporal patterns, i.e. acute, acute recurrent, chronic progressive/non-progressive, episodic or constant.

DIAGNOSTIC CRITERIA

» Exclude secondary causes of headache, e.g. raised intracranial pressure.
» Red flags in childhood headaches:
  > change in pattern (e.g. 'worst headache ever'),
  > progressive course over time,
  > age younger than 3 years,
  > nocturnal/wakes child from sleep,
  > early morning vomiting,
  > ataxia,
  > focal neurological signs,
  > alteration of level of consciousness.

GENERAL AND SUPPORTIVE MEASURES

» Environmental and lifestyle changes, e.g. avoid precipitants such as bright lights, sleep deprivation and certain foods, excessive video games.
» Adequate hydration.
» Avoid skipping meals, excessive caffeine ingestion.
» Regular exercise.
» Stress alleviation and coping skills training where possible.
» Headache diary and identify possible triggers.

MEDICINE TREATMENT

Treat non-migraine headaches with analgesics.

- Paracetamol, oral, 15 mg/kg/dose 6 hourly as required.

For migraine:

- Ibuprofen, oral, 10 mg/kg/dose, 6 hourly.

Persistent vomiting and not tolerating oral feeds:

- Metoclopramide, oral, 0.15–0.3 mg/kg as a single dose.

OR

- Metoclopramide, IM/IV, 0.1 mg/kg as a single dose.

OR

- Ondansetron, oral, 0.1–0.2 mg/kg 12 hourly.

Note:
Headaches can be an adverse effect associated with the of use ondansetron. Patients with ongoing symptoms should be investigated.

Migraine prophylaxis
Indicated when headaches occur frequently, impacting on the child's activity and requiring substantial relief medication.
Treat for six months then review.

- Propranolol, oral, 0.5–3 mg/kg/day in 2–3 divided doses.
  - Contraindicated in asthma and heart block.
  - Avoid in diabetes and depression.

In children who are unable to take propranolol, e.g. asthma:

- Topiramate, oral, 1–3 mg/kg/day in 1–2 doses (specialist initiated).
  - Starting dose: 0.5 mg/kg/day.
  - Titrate dose slowly every 1–2 weeks.
  - Reinforce behavioural management before considering topiramate.

REFERRAL

» Secondary intracranial cause suspected.
» Failure to respond to first line treatment.

13.6 NEUROCYSTICERCOSIS

B69.0

DESCRIPTION

Neurocysticercosis is caused by the cysticercal form, i.e. larval form, of the pork tapeworm, *Taenia solium*. The larvae may locate in the brain parenchyma, intraventricular and meningeal areas, spinal canal/cord and eye, or a combination of these regions. Viable cysticerci incite little inflammatory response, but dead cysticerci elicit an increased inflammatory response.
Cysticerci in the brain may remain dormant or may cause complications such as:

» headache,
» behavioural disorders,
» focal neurological deficits,
» raised intracranial pressure,

» visual disturbances,
» seizures,
» meningo-encephalitis,
» hydrocephalus,
» meningitis,
» spinal cord compression.

DIAGNOSTIC CRITERIA

Clinical

» Location and stage of the life cycle of the parasite in the brain determines the clinical features.
» Suspect if child from an endemic area, i.e. pig farming area, presents with neurological abnormalities such as:
  > seizures,
  > raised intracranial pressure/hydrocephalus,
  > focal neurological deficits,
  > cranial nerve palsies.
  > meningo-encephalitis,
  > meningitis,
  > behavioural disorders,
  > headache,

Investigations

» Computed tomography (CT scan) and/or magnetic resonance imaging (MRI scan) of brain showing cysts, granulomas, peri-lesional oedema or calcification of cysts.
» MRI scan may identify more lesions and viable cystic lesions than the CT scan.
» Soft tissue radiology of muscles of lower limbs may demonstrate calcified cysticerci, i.e. 'rice grain' calcifications in muscles.
» Follow-up CT scans and/or MRI scans may help to assess the response to therapy.

GENERAL AND SUPPORTIVE MEASURES

Prevention:

» Prolonged freezing or thorough cooking of pork to kill the parasite.
» Thorough washing of fresh fruit and vegetables in *T. solium* endemic areas.
» Attention to personal hygiene after use of toilet.
» Proper sanitation facilities and safe water.
» Avoid the use of human excreta as fertiliser.
» Look for Taenia ova in the stools of the family members.

MEDICINE TREATMENT

Calcified cysticerci and a single dying lesion visible on CT scan require no anti-helminthic treatment.

Patients with multiple cysts usually have a mixture of live and dying cysts and are assumed to have active disease and require treatment.

- Albendazole, oral, 7.5 mg/kg/dose 12 hourly for 7 days.
  - Maximum dose: 400 mg/dose.

Prevention of neurological manifestations

In massive infestations, cysticidal therapy may trigger an inflammatory response. Delaying anti-helminthic therapy and adding corticosteroids may lessen the risk.

24 hours **prior** to albendazole therapy:

- Dexamethasone, IM, 0.15 mg/kg/dose 6 hourly.

Then follow with oral therapy as soon as possible:

- Prednisone 1 mg/kg/day for the duration of albendazole therapy, and then taper and discontinue.

Seizure control

See section 13.4: Epilepsy.
Treat according to the type of seizure.
AED treatment for 6–12 months after resolution of lesions on neuroimaging.
Recurrent seizures require chronic treatment until seizure-free for 2 years.

REFERRAL

» Neurocysticercosis not responding to adequate therapy.
» Neurocysticercosis with complications, such as hydrocephalus.
» Intractable epilepsy.

13.7 NEUROMUSCULAR DISORDERS

13.7.1 INFLAMMATORY POLYNEUROPATHY (GUILLAIN-BARRÉ SYNDROME) *

G61.0
* Notifiable condition

DESCRIPTION

Guillain-Barré syndrome (GBS) is an acute autoimmune-mediated polyradiculoneuropathy which is precipitated by a preceding viral or other infection. It is the most common acquired polyneuropathy in children.

Different forms or **variants of** Guillain-Barré syndrome are described depending on the clinical and/or neurophysiological characteristics.

Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP)

» This is the most common form, accounting for 80–90% of cases.
» Characterised mainly by:
  > symmetrical, ascending motor weakness,
  > areflexia, i.e. absence of tendon reflexes,
  > distal sensory alteration,
  > pain/paraesthesia.

Acute Motor Axonal Neuropathy (**AMAN**)

» A purely motor form of GBS.
» It involves predominantly motor nerves and has an axonal pattern on electrophysiology (nerve conduction studies).
» Although there are similarities with AIDP, the clinical picture tends to be more severe with more patients suffering from respiratory failure.

Acute Motor-Sensory Axonal Neuropathy (**AMSAN)**
» Another axonal form of GBS but with sensory involvement.
» It is not frequently found in children.

Miller-Fisher syndrome
» Patients have external ophthalmoplegia, sensory ataxia, weakness with areflexia.
» Electrophysiological and CSF studies are similar to AIDP.

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (**CIDP**)
» May be considered a chronic variant of AIDP.
» Most often starts insidiously and progresses slowly but can have onset like GBS.
» It is managed differently from GBS and should be referred.

DIAGNOSTIC CRITERIA

Clinical
» Preceding respiratory tract or gastrointestinal infection.
» Symmetrical, flaccid muscle weakness with early areflexia.
» The muscle weakness may ascend rapidly upwards to involve the trunk, arms, face and cranial nerves.
» Bulbar paralysis and respiratory failure may develop.
» Autonomic dysfunction.
» Relatively mild, or absence of, sensory signs.
» Exclude the following:
  > Acute Disseminated Encephalomyelitis (ADEM),
  > poliomyelitis, a rare cause of hypotonia with abrupt onset of weakness (usually asymmetrical) in association with a febrile illness,
  > transverse myelitis:
    - initial flaccid weakness and areflexia typically involving the lower limbs maximally,
    - occasionally with pain at the onset, but rapidly progressing to spasticity and hyperreflexia,
    - a sensory level on the trunk,
    - bladder and rectal sphincter involvement.
  > diphtheria,
  > botulism.

Investigations
Follow the Acute Flaccid Paralysis (AFP) investigation protocol

» Send two stool specimens taken 24–48 hours apart to the National Institute of Virology via the local laboratory.
» The stool sample needs to be sent within 14 days of onset of paralysis to exclude poliovirus infection.

CSF

» CSF findings after 1 week show elevated protein and few or no cells, i.e. albumino-cytological dissociation.
» CSF glucose is normal.

GENERAL AND SUPPORTIVE MEASURES

» Notify as AFP.
» Admit to a high care or intensive care unit.
» Monitor respiratory and autonomic functions closely:
  > peak expiratory flow rate,
  > respiratory rate,
  > forced vital capacity (FVC),
  > arterial blood gases.
  > blood pressure,
  > heart rate,
  > bulbar functions,
» Ventilation is recommended when:
  > rapidly progressing ascending paralysis, including shoulder weakness, head lag, weak cough and swallowing difficulties,
  > there is a progressive fall in the peak expiratory flow rate,
  > tachycardia and sweating occur,
  > inspiratory efforts are weak (typical signs of respiratory distress will be absent),
  > inability to talk,
  > P_aCO_2 levels start rising.

  Note:
  These changes precede hypoxaemia detected on blood gas analysis, and ventilation should begin before frank hypoxaemia occurs.
  Respiratory care must be meticulous.

» To determine fluid losses from autonomic instability, monitor urine output and degree of sweating.
» Provide adequate nutrition.
» Provide bladder and bowel care as well as pressure-point care.
» Routine physiotherapy for chest and limbs, keep ankles in neutral position (90°) (may require foot/hand splints).
» Protect eyes and keep moist.
» Communicate with child as awareness is maintained. Staff should remember that children may be very frightened but unable to express their emotions and needs.

MEDICINE TREATMENT

- Immunoglobulin, IV, 1 g/kg/day, slowly over 12–16 hours on two consecutive days **or** 0.4 g/kg as a single daily dose on 5 consecutive days early in the disease process.
  - Use under specialist supervision.

Substantial pain is present (in up to 90%) in the severely affected patients. Pain in this setting is often unrecognised and underestimated.
Pain management is essential. See section 20.1.1: Management of pain.

For neuropathic pain**:**

- Carbamazepine, oral, 5 mg/kg/dose 12 hourly.

REFERRAL

» Chronic inflammatory demyelinating polyradiculoneuropathy.
» Guillain-Barré syndrome with bulbar paralysis and/or early signs of respiratory failure.
» Patients who have lost or are losing ambulation for management in consultation with a paediatric neurologist.
» Patients with complex Guillain-Barré syndrome.

13.7.2 MYASTHENIA GRAVIS

G70.0

DESCRIPTION

An auto-immune disorder resulting in muscle fatigue. Mild cases involve the eyes alone, i.e. ptosis and ophthalmoplegia, and severe cases involve proximal muscle groups, respiratory and bulbar control.

DIAGNOSTIC CRITERIA

Clinical

» Muscle fatigability with exercise and demonstration of this in the clinic setting:
  > Lid-lag test, i.e. failure to maintain upward gaze for 1 minute.
  > Arm-raising test, i.e. failure to maintain the arms at 90° from the trunk for 1 minute.

Note:
Myasthenia gravis patients not uncommonly present in a myasthenic crisis, with bulbar and respiratory compromise. Sometimes this may be the first mode of clinical presentation.

MEDICINE TREATMENT

- Pyridostigmine, oral, 1–5 mg/kg/day in 4–6 divided doses. (Specialist initiated).

REFERRAL

» All for confirmation of diagnosis and initiation of treatment (consideration of steroids, immuno-modulation therapy).
» Myasthenic crisis.

13.7.2.1 MYASTHENIC CRISIS (MC)

G70.01

DESCRIPTION

Acute onset of respiratory failure due to worsening myasthenia gravis, requiring ICU admission. Respiratory and bulbar insufficiency is common with an inability to breathe or swallow or may have worsening of existing symptoms. MC is most frequently precipitated by systemic infections.

Myasthenic crisis may be the initial presentation.

DIAGNOSTIC CRITERIA

Clinical

» Worsening of existing weakness.
» Respiratory compromise.
» Inability to swallow.

GENERAL AND SUPPORTIVE MEASURES

» Admit to ICU for close observation.
» Provide ventilatory and feeding support as required.

MEDICINE TREATMENT

For consideration of glucocorticoid therapy and intravenous immunoglobulin, in discussion with neurologist.

- Dexamethasone, IV or IM,

- Immunoglobulin, IV, 1 g/kg/day, slowly over 12–16 hours on two consecutive days **or** 0.4 g/kg as a single daily dose on 5 consecutive days early in the disease process.

REFERRAL

» All cases for further management.

13.7.3 DUCHENNE MUSCULAR DYSTROPHY (DMD)

G71.01

» An X-linked recessive disorder causing progressive muscle weakness, typically in males.
» Carrier females may have some degree of weakness.
» Due to a mutation in the gene coding for dystrophin.
» There may be a family history of DMD.

DIAGNOSTIC CRITERIA

Clinical

» Delayed walking.
» Toe walking.
» Gowers sign.
» Waddling gait.
» Lumbar lordosis.
» Calf pseudohypertrophy.
» Short stature.
» Progressive proximal muscle weakness, usually confined to a wheelchair by age 13 years.
» Cardiomyopathy
» May have mild cognitive impairment.
» Behavioural issues.

Investigations

» CK markedly elevated.
» AST/ALT may be elevated.

GENERAL AND SUPPORTIVE MEASURES

» Pain may be present and should be treated appropriately.
» Physiotherapy
» Occupational therapy.
» Nutritional support.
» Encourage gentle aerobic exercise.
» Psychosocial support for patient and family.

MEDICINE TREATMENT

Consider oral corticosteroids once plateau or decline in motor function, in consultation with neurologist.

- Prednisone, oral, 0.75 mg/kg daily.
    - Maximum dose 30–40 mg.
- Long term use of corticosteroids is associated with various complications. Monitor patients and manage as needed.

If pain is present, refer to Chapter 20: Pain Control, section 20.1.1: Management of pain.

REFERRAL

All cases for specialist assessment.

13.8 ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM)

G04.0

DESCRIPTION

» Most common demyelinating disorder in childhood.
» Affects children with a peak incidence between 5 and 8 years.
» May occur following a systemic viral illness or vaccination.
» Demyelination of white matter in multiple areas of the brain and spinal cord.

DIAGNOSTIC CRITERIA

» ADEM is a diagnosis of exclusion.
» Consider and exclude differential diagnoses: SLE, infectious or autoimmune encephalitis, metabolic disorders, hypertensive encephalopathy.

Characterised by acute onset of encephalopathy with focal or multifocal neurological deficits.

» Seizures
» Cranial nerve palsies.
» Meningism
» Optic neuritis.
» Gait disturbances.
» Hemiparesis
» Pyramidal signs.
» Ataxia
» Aphasia

Symptoms usually peak between 2 and 5 days from onset but may change or worsen for up to 3 months.

CSF

» Normal pressure.
» May have mild pleocytosis.
» Increased protein.
» Normal glucose.

Diagnostic criteria:

1. A first, polyfocal clinical CNS event with a presumed inflammatory demyelinating cause.
2. Encephalopathy (alteration in consciousness or behavior unexplained by fever, systemic illness or postictal syndrome).
3. Brain MRI abnormalities consistent with demyelination during the acute (initial three-month) phase.
4. No new clinical or MRI findings three months or more after clinical onset.

GENERAL AND SUPPORTIVE MEASURES

» Supportive care including ICU and mechanical ventilation may be necessary.

MEDICINE TREATMENT

Consider treatment with immune modulators and immunoglobulins in consultation with a neurologist.

REFERRAL

» As MRI is required for diagnosis, all patients should be referred prior to implementing treatment.

13.9 SYDENHAM CHOREA

I02.9

DESCRIPTION

A movement disorder with rapid involuntary jerks affecting any part of the body often incorporated into a voluntary movement in an attempt to mask it. It is an acute post-streptococcal infection movement disorder and constitutes one of the major criteria for the diagnosis of rheumatic fever. Patient has the appearance of being restless with constant movement which improves with sleep. The movements are classically random in place and random in time.

DIAGNOSTIC CRITERIA

Clinical

» Exclude drug reactions, hyperthyroidism, systemic lupus erythematosus and neurodegenerative disorders.

Investigations

» Cardiac screening, i.e. ECG, echocardiogram.
» Serum ASOT, anti-DNAse B.
» Erythrocyte sedimentation rate.
» Anti-dsDNA, if clinically indicated.

GENERAL AND SUPPORTIVE MEASURES

» Emotional support.
» School support.
» Occupational therapy.

MEDICINE TREATMENT

Movement disorders:

- Haloperidol, oral, 0.025 mg/kg/day in 2–3 divided doses.
  - Increase dose slowly and incrementally to 0.05 mg/kg/day.

PLUS

If streptococcal infection:

- Phenoxymethylpenicillin, oral, 500 mg 12 hourly for 10 days.

OR

- Amoxicillin, oral, 45 mg/kg/dose 12 hourly for 10 days.

THEN

Until 21 years of age:

- Benzathine benzylpenicillin (depot formulation), IM, 1.2 million units every 28 days.

OR

- Phenoxymethylpenicillin, oral, 250 mg 12 hourly.

REFERRAL

» All patients for specialist assessment.

13.10 CEREBROVASCULAR DISEASE/STROKE

I67.9

DESCRIPTION

Cerebrovascular disease can be ischaemic (thrombotic or embolic) or haemorrhagic, arterial or venous. Arterial ischaemic stroke must always be considered in any child with sudden onset of hemiparesis or other focal neurological disturbance. The clinical features of cerebral venous thrombosis (CVT) include headache, papilloedema, focal neurological signs, seizures (often focal), and alteration of consciousness.

Risk factors:

» cardiac disorders,
» infections, e.g. meningitis, varicella, HIV, etc.,
» prothrombotic disorders, e.g. nephrotic syndrome, protein S/C deficiencies, etc.,
» haematologic disorders, e.g. sickle cell anaemia,
» vasculopathies, e.g. vasculitis, HIV, Moyamoya syndrome.

The initial evaluation in children includes the following:

CT/MRI brain to ascertain whether it is an ischaemic or haemorrhagic infarct.

» Electrocardiography, echocardiography.
» Full blood count, INR, PTT.
» CSF analysis as indicated.
» Infectious screening, including varicella, HIV, mycoplasma, TB.
» Connective tissue and vasculitic screening.
» Thrombophilia screening. See Chapter 3: Blood and Blood Forming Organs, section 3.12: Venous thrombo-embolic disease.

GENERAL AND SUPPORTIVE MEASURES

Acute supportive and neuroprotective care directed at preserving damaged but salvageable brain tissue includes the following:

» Maintain body temperature in the low to normal range.
» Maintain euglycaemia.
» Maintain O_2 saturation above 95%.
» Maintain adequate cerebral perfusion and manage raised intracranial pressure.
» Treat anaemia.
» Treat acute seizures promptly.

Haemorrhagic stroke requires referral to a centre with neurosurgical expertise and facilities.
Early disability assessment and management, includes physiotherapy, speech therapy, occupational therapy, etc.

MEDICINE TREATMENT

Arterial ischaemic stroke without haemorrhage

All patients with confirmed arterial ischaemic stroke:

- Aspirin soluble, oral, 1–5 mg/kg as a daily dose.
  - Contraindicated in haemorrhagic stroke or bleeding tendency.

REFERRAL

» All patients to specialist paediatrician for investigation.
» Anticoagulation with enoxaparin and warfarin is best done in a specialised setting under cardiologist, haematologist and neurologist supervision.

13.11 LUMBAR PUNCTURE

CONTRAINDICATIONS TO LUMBAR PUNCTURE

» Focal neurological signs and depressed level of consciousness.
» Clinical signs of raised intracranial pressure, or impending cerebral herniation:
  > deep coma, i.e. GCS < 9, or sudden deterioration of level of consciousness,
  > decerebrate or decorticate posturing,
  > neurogenic hyperventilation,
  > unequal dilated or poorly reactive pupils,
  > absent doll's eye reflex,
  > papilloedema.
» Haemodynamic/respiratory unstable patients.
» Clinical meningococcaemia (septicaemia) with petechiae/purpura. (confirm with skin scrape, Gram stain and blood culture).
» Skin sepsis or abnormalities over the lumbar puncture site.
» Coagulopathy

» Spinal anatomic abnormality.
» Acute paraplegia.
» Status epilepticus.

PROCEDURE

» Positioning and restraint are vital in determining the success of the procedure.
» The ability of the assistant in restraining is as important as the skill of the 'operator'.
» Preparation entails not only positioning, but attention to sedation/analgesia, 'patient comfort' and safety, as well as factors such as adequate lighting.
» Resuscitation equipment must be available at the bed side.
» Pay attention to the sterility of the operating field.
» Local analgesia with/without sedation may be required. See Chapter 20: Pain Control, section 20.1.1: Management of pain.
» Ensure that all necessary equipment, e.g. needles, manometers and specimen tubes are close at hand.
» Only the interspaces below L3 (L3/L4 or L4/L5) are used in order to avoid damaging the conus medullaris.
» With the patient in the lateral recumbent position, the L3/L4 interspace is found at the level of the line joining the highest points of the two iliac crests.
» Turn the bevel of the needle (with stylet) to face the patient's side to avoid cutting the longitudinal dural fibres.
» As the needle is advanced, the first 'give' or loss of resistance is encountered with the piercing of the ligamentum flavum. A slight 'popping' sensation is felt as the needle penetrates the dura. Remove the stylet to allow CSF to drain out passively. If no fluid appears, then rotate the needle a quarter turn (90°). If this does not help, replace the stylet and advance the needle a few millimetres and then check for fluid as before.
» Measure the opening pressure using a manometer, with the child relaxed in the lateral decubitus position. In a young, relaxed child, the opening pressure is in the range of 6-18 cm H_2O.
» At the end of the procedure, re-insert the stylet before removing the needle completely.

Note:
If intracranial infection is suspected, do a blood culture and initiate antimicrobial treatment immediately. See Chapter 8: Infective/Infectious Diseases, section 8.11: Meningitis, Acute Bacterial.

Remember to catch a few drops of CSF on a labstick to check the glucose and for the presence of white cells which may give an indication of an infection.

13.12 RAISED INTRACRANIAL PRESSURE

DESCRIPTION

Raised intracranial pressure (ICP) is an emergency requiring prompt recognition and treatment.

The cranial vault contains the brain, blood and CSF. It has a fixed volume, therefore, an increase in one component requires a compensatory decrease in others to maintain the pressure within the compartment. There is a limited capacity for compensation, i.e. by decreasing CSF volume, decreasing cerebral blood volume or by increasing the cranial volume. Thereafter, there is a rise in pressure.

Herniation syndromes are an important complication to consider. They may arise in the course of untreated underlying disease or as a result of injudicious lumbar puncture.

Normal CSF pressures range from:

- Infants: 20 – 27 cmH_2O (1.5–6 mmHg)
- Children: 4 – 10 cmH_2O (3–7 mmHg)

These values are for well children and may vary in those who are critically ill on ventilation.
Any CSF pressure above 27 cm of CSF (20 mmHg) for more than 20 minutes should be treated.

Increased brain volume	Intracranial space occupying lesion: Brain tumour, brain abscess, haematoma, vascular malformation, arachnoid/ epidermoid cyst.
	Cerebral oedema: encephalitis, meningitis, hypoxic ischaemic encephalopathy, traumatic brain injury, hepatic encephalopathy, Reye's syndrome, stroke, diabetic ketoacidosis, hyponatraemia.
Increased blood volume	Vascular malformations, cerebral venous thrombosis, meningitis, encephalitis.
Increased CSF volume	Obstructive/communicating hydrocephalus, choroid plexus papilloma.
Disordered CSF dynamics	Idiopathic intracranial hypertension.

Source[20]

Vitamin A administration may cause benign raised ICP.

DIAGNOSTIC CRITERIA

Presentation may have an acute or insidious onset depending on the underlying pathology. A detailed history and examination are essential.

Clinical

Features vary with age.

Infants:

- Increasing head circumference.
- Sun setting eyes.
- Distended scalp veins.
- Irritability
- Lethargy
- Vomiting
- Developmental delay or regression.
- Persistent head lag.

Older children:

- Headache
- Vomiting
- Depressed level of consciousness.
- Seizures
- Ataxia
- Abnormal eye movements.
- Double vision.
- Behavioural changes.
- Meningism

Late signs:

- Papilloedema
- Sixth nerve palsy.
- Pupillary dilation.
- Decerebrate or decorticate posturing.
- Cheyne-Stokes respiration.
- Focal neurological deficits.
- Cushing Triad:
  - Increased systolic pressure (with widening pulse pressure).
  - Irregular breathing.
  - Bradycardia

Herniation syndromes	
Unilateral transtentorial herniation	Declining consciousness. Increased blood pressure. Slow pulse. Homonymous hemianopia. Respiratory irregularity.
Bilateral transtentorial herniation	Decerebrate or decorticate rigidity. Declining consciousness. Impaired upward gaze. Irregular respiration. Pupillary constriction or dilatation.
Cerebellar herniation	Declining consciousness. Impaired upward gaze. Irregular respiration. Lower cranial nerve palsies. Neck stiffness or head tilt.

Source[20]

Investigations

» CT brain to determine underlying cause and whether it is safe to perform a lumbar puncture.
» CSF opening pressure (if no contraindication).

GENERAL AND SUPPORTIVE MEASURES

» Follow the ABCD algorithm.
» Position head in midline and elevate to 30°.
» Cautious ventilation maintaining P_aCO_2 between 4.5 and 5 kPa.
» Monitoring and maintenance of blood pressure
» Elevated BP is usually reactive and required to maintain cerebral perfusion.
» Monitor fluid balance, use 0.9% NaCl/5% dextrose water as maintenance fluid.
» Keep serum sodium in the upper range of normal, up to 150 mmol/L.
» Maintain normothermia.
» Maintain glucose within 6 to 10 mmol/L.

MEDICINE TREATMENT

Initiate treatment for underlying cause.
If meningitis cannot be excluded, commence treatment as soon as possible (see Chapter 8: Infective/Infectious Diseases, section 8.11: Meningitis, acute bacterial).

Sodium chloride, 5%, IV, 2 mL/kg infused over 30 minutes.

- Monitoring of serum sodium is essential with repeat doses or infusion.

OR

Mannitol, IV, 250 mg/kg administered over 30–60 minutes.

- Do not exceed two doses without consulting with a specialist.

If a space occupying lesion is diagnosed:
Add

- Dexamethasone, IV, 0.5 mg/kg 12 hourly.
- Maximum dose 12 mg per dose.

Sedation and analgesia, see Pain Control Chapter and Intensive Care Chapter.
Management of seizures if present, refer to section 13.1: Seizures.

REFERRAL

» According to underlying condition.
» Neurosurgical intervention may be required.

13.13 CEREBRAL PALSY (CP)

ICD 10 G80.9

DESCRIPTION

Cerebral palsy (CP) describes a group of disorders of movement and posture. It is the commonest cause of developmental disturbances in children. CP results from an insult to the developing foetal or infant brain, which is non-progressive. There may be associated abnormalities of sensation, perception, cognition, communication and behaviour.

DIAGNOSTIC CRITERIA

» Motor deficit with delay in motor milestones.
  > Not sitting by 8 months (corrected for gestational age).
  > Not walking by 18 months (corrected for gestational age).
  > Hand preference before 12 months (corrected for gestational age).
» No loss of function (i.e. milestone regression).
» Serial examinations may be needed to establish the diagnosis.

Clinical picture may be described by:

» The predominant abnormality of tone and the distribution of areas involved.

Regional involvement	Spastic	Hemiplegia
	Spastic	Diplegia
Global involvement	Spastic	Quadriplegia
	Dyskinetic	Athetoid
	Dyskinetic	Dystonic
	Ataxic	Ataxia

Risk factors:

» Preterm labour and birth.
» Small for gestational age.
» Multiple pregnancy.
» Neonatal encephalopathy.
» Neonatal sepsis.
» Meningitis or septicaemia.

Developmental follow up from birth to 2 years is essential in high-risk patients.

GENERAL AND SUPPORTIVE MEASURES

» Multidisciplinary team approach.
» Screen for ophthalmologic and hearing impairments.
» Screen for speech and language disorders.
» Monitor growth, nutrition and swallowing function.
» Physiotherapy
» Occupational therapy.
» Psychology
» Assess level of functioning according to the Gross Motor Function Classification System (GMFCS).

MEDICINE TREATMENT

Medication for relief of spasticity, dystonia and dyskinesia may be used in some patients for improvement in function, range of motion at various joints, disturbances from uncontrolled movements.

Co-morbidities and complications:

Neurological	Epilepsy, hydrocephalus, visual and hearing impairment.	See section 13.4: Epilepsy.
Behavioural and learning	Sleep disturbance, depression, autistic features, learning difficulties, vulnerability.	See Chapter 14: Child and Adolescent Psychiatry, sections 14.4.1: Depression, 14.12: Autism spectrum disorders.
Gastro-intestinal	Difficulty swallowing, gastro-oesophageal reflux, constipation.	See Chapter 2: Alimentary Tract, sections 2.2.2: Constipation and 2.2.8: Gastro-oesophageal reflux disease (GORD).
Bone	Osteoporosis, scoliosis, hip dislocation, pathological fractures.	
Respiratory	Susceptibility to chest infections and aspiration.	See Chapter 15: Respiratory System.
Skin	Drooling, pressure sores.	
Dental	Poor oral hygiene, susceptibility to dental caries.	See Primary Health Care STGs and EML, Chapter 1 Dental and oral conditions, section 1.1: Abscess and caries.
Genito-urinary	Neurogenic bladder	See Chapter 6: Nephrological/Urological disorders, Dysfunctional bladder.

Table adapted from: Smith, M and Kurian, M. Medical management of cerebral palsy. Paediatrics and Child Health, 2016-09-01, Volume 26, Issue 9, Pages 378–382.

These patients frequently experience pain as a result of these complications.
Remember to suspect and manage pain in children with CP.

REFERRAL

For consideration of medical treatment of spasticity, dystonia or dyskinesia in consultation with the multidisciplinary team.

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