

What is a syndrome?

A syndrome is a group of signs or symptoms that happen together and help to identify a unique medical condition. Epilepsy syndromes are usually diagnosed in childhood but some can continue into adult life.

What is a childhood epilepsy syndrome?

If your child is diagnosed with an epilepsy syndrome, it means that their epilepsy has some specific signs and symptoms. These include:

- the type of seizure or seizures they have;
- the age when the seizures start;
- a specific pattern on an electroencephalogram (EEG); and
- sometimes a specific pattern on a brain imaging scan.

An EEG test is painless and records patterns of electrical activity in the brain. Some epilepsy syndromes have a particular pattern, so the EEG can be helpful in finding the correct diagnosis. A magnetic resonance imaging (MRI) brain scan is also painless and looks at the structure of the brain for any underlying abnormality.

An epilepsy syndrome can only be diagnosed by looking at all the signs and symptoms together.

Visit epilepsysociety.org.uk/diagnosis

If your child is diagnosed with an epilepsy syndrome, it may help the paediatrician (a doctor who specialises in treating children) to plan their care (for example, choosing treatment options or deciding whether further tests are needed).

Different types of syndrome

Syndromes can vary greatly. Some are called 'benign' which means children become seizure-free (have no seizures) once they reach a certain age. Other syndromes are 'severe' and children have seizures which are difficult to control. Anti-seizure medications (ASMs) may be tried alone, or in combination with each other, and some non-drug treatments may also be tried, for example the ketogenic diet.

Many children with severe epilepsy syndromes have additional difficulties with learning and behaviour and may need extra support.

Examples of childhood syndromes

Benign rolandic epilepsy (BRE)

This syndrome affects 15% of children with epilepsy and can start any time between the ages of 3 and 10.

Children may have very few seizures and most become seizure-free by the age of 16. They may have focal motor aware seizures, which means they have movement involving their face or limbs without losing awareness. They happen mostly on falling asleep, on waking up, or during sleep, and usually involve the muscles that involve speech and swallowing, causing gurgling or grunting noises, mouth movements, and dribbling. Speech can be temporarily affected and symptoms may develop into a tonic clonic seizure. ASMs may not be necessary, but can be helpful if seizures are more frequent or are mostly tonic clonic.

Childhood absence epilepsy (CAE)

This syndrome starts between the ages of 4 and 10, and can affect up to 12% of children with epilepsy under 16. Absence seizures happen on daily basis (up to 100 times a day) and are very brief, lasting only a few seconds. Because of this, they often go unnoticed.

During a seizure a child will become unconscious. They may look blank or stare, their eyelids may flutter, and they may make repetitive movements. They may not respond to what is happening around them, or be aware of what they are doing. Seizures respond well to medication. If a child is seizure-free for two years, medication is sometimes gradually reduced. Up to 90% of children with CAE will grow out of seizures by the time they are adolescents. Sometimes a child may also have other types of seizures.

Dravet syndrome

Dravet syndrome is a rare and complex type of epilepsy that affects around one in every 15,000 people in the UK. Seizures usually begin in the first year of life, with additional characteristics emerging typically from the second year onwards.

There are many different types of epilepsy syndrome. This factsheet gives a brief overview of what epilepsy syndromes are and includes details of some specific syndromes and their characteristics.

Helpline 01494 601400
Confidential, national call rate.
Information and emotional support.
Visit epilepsysociety.org.uk/helpline
for opening hours.

The seizures occur spontaneously and may often also be associated with a high temperature or a hot environment.

This syndrome is often associated with difficulty to treat seizures, intellectual and behavioural difficulties, and a range of other problems. Dravet syndrome is lifelong, though different aspects may emerge or change as time goes on.

Juvenile myoclonic epilepsy (JME)

This syndrome usually starts between the ages of 12 and 18. Many children have different types of seizure: myoclonic seizures (brief muscle jerks) in the upper body, and tonic clonic seizures. Some children may also experience occasional brief absence seizures.

Seizures often happen as, or shortly after, the child or young person wakes up. Medication can be successful in controlling seizures, and may be needed for life. Tiredness, stress, lack of sleep, and excess alcohol can trigger seizures. Up to 40% of children or young people with JME have photosensitivity, where seizures are triggered by flashing lights or contrasting light or dark patterns.

[Visit epilepsysociety.org.uk/photosensitive-epilepsy](https://www.epilepsysociety.org.uk/photosensitive-epilepsy)

Infantile spasms (or West Syndrome)

This syndrome often begins in the first year of life and can affect children:

- who have had a previous brain injury before the age of six months;
- whose brain has not formed properly (brain malformation);
- who have genetic abnormalities.

It is identified by brief spasms or jerks which happen in 'clusters'. Spasms can affect the whole body or just the arms and legs. Each cluster can include between 10 – 100 individual spasms, which often happen when the child is waking up. ASMs and corticosteroids (medicines to reduce inflammation) are used to treat this syndrome, although around 25% of children have spasms that do not respond well to medication. Many children develop problems with learning or behaviour. Some may go on to develop Lennox-Gastaut syndrome.

Lennox-Gastaut syndrome

This syndrome usually begins between the ages of 3 and 5, but can start as late as adolescence. Children may have different types of seizures, most commonly tonic (where muscles become stiff), atonic (where muscles relax), and atypical absences.

Every effort is made to ensure that all information is correct at the time of printing. Please note that information is intended for a UK audience. This information is not a substitute for advice from your own doctors. Epilepsy Society is not responsible for any actions taken as a result of using this information.

Atypical absences are different from typical absences as they often last longer, and the pattern on an EEG is more irregular.

Many children also develop learning difficulties and behaviour problems. This syndrome can be very difficult to treat with ASM, and most children need a combination of different drugs. Some non-drug treatments, such as the ketogenic diet and vagus nerve stimulation therapy (VNS), can also be helpful. Seizures often continue into adult life.

[Visit epilepsysociety.org.uk/treatment](https://www.epilepsysociety.org.uk/treatment)

Who can I talk to?

If your child has been diagnosed with a childhood epilepsy syndrome, you may have concerns or questions. You can get information and advice from a paediatrician with an interest in epilepsy or a paediatric neurologist, or from a neurologist for adults. Support may also be available through an epilepsy specialist nurse, counsellor, support group, friends, family, or a helpline. Epilepsy Society has a confidential helpline that offers time to talk, information, and emotional support (see page 1).

Further reading and support

[corpal.org.uk](https://www.corpal.org.uk) - supporting those with ACC or Aicardi Syndrome.

[angelmanuk.org](https://www.angelmanuk.org) - Angelman Syndrome support, education, and research trust.

[dravet.org.uk](https://www.dravet.org.uk) - support, education, and research for those affected by Dravet Syndrome.

[lgsfoundation.org](https://www.lgsfoundation.org) - Support for Lennox Gastaut syndrome in the UK.

[rettsyndrome.org.uk](https://www.rettsyndrome.org.uk) - information on Rett Syndrome.

[ring20researchsupport.co.uk](https://www.ring20researchsupport.co.uk) - research, information, and support for Ring Chromosome 20 Syndrome.

[sturgeweber.org.uk](https://www.sturgeweber.org.uk) - Sturge Weber Foundation

[wssg.org.uk](https://www.wssg.org.uk) - West Syndrome information

[contact.org.uk](https://www.contact.org.uk) - Contact a family. Information and links to support for many childhood conditions, including epilepsy syndromes.

For a printed copy of this information contact our helpline.

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