Epilepsy syndromes

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 an '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 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. Children may need a general anaesthetic to ensure they remain still during the MRI scan.

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

Visit epilepsysociety.org.uk/diagnosis and epilepsysociety.org.uk/epileptic-seizures

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' or 'self-limited', 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.

Visit epilepsysociety.org.uk/treatment

Examples of childhood syndromes

Benign rolandic epilepsy (BRE)

BRE affects around 15% of children with epilepsy and can start any time between the ages of 3 and 13.

Children may have very few seizures and most become seizure-free by the age of 16. They may have focal preserved consciousness seizures with observable manifestations (previously called focal motor aware seizures), which means they involve movement. They are often at night, and usually involve one side of the face and/or 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 usually starts between the ages of four and ten, and can affect up to 18% of school age children with epilepsy.

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 0300 102 0024
Confidential, information, and emotional support.
Visit epilepsysociety.org.uk/helpline for opening hours.

Absence seizures happen frequently (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 anti-seizure 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 adults. Sometimes a child with CAE may also have other types of seizures.

Dravet syndrome

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

The seizures occur spontaneously and may often be associated with a high temperature or a hot environment.This syndrome is often associated with difficult 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.

For information about Epilepsy Society research visit epilepsysociety.org.uk/news/epilepsy-society-research-published-brain-communications-journal and epilepsysociety.org.uk/news/treatment-ultra-rare-form-epilepsy-shows-promising-results

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 generalised 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. It is common for people with JME to have photosensitive epilepsy.

Visit 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); or
- 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, and medicines help to control spasms in about seven out of ten children. 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 three and five, 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.

Atypical absences are different from typical absences as they often last longer, and a child may be responsive and aware of their surroundings.

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 and epilepsysociety.org.uk/news/life-changing-treatment-lennox-gastaut-syndrome

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 (see page 1).

Further information and support

Angelman Syndrome
angelmanuk.org
Angelman Syndrome support education and research trust (ASSERT).

Cambridge Rare Disease Network UK
camraredisease.org
Unites patients, advocates, experts and leaders to address the challenges faced by people affected by rare diseases.

Contact a family
contact.org.uk
Information and links to support for many childhood conditions, including epilepsy syndromes.

Corpal
corpal.org.uk
Supporting those with disorders of the corpus callosum (DCC) or Aicardi Syndrome.

Dravet Syndrome UK
dravet.org.uk
Support education and research for those affected by Dravet Syndrome.

USA Epilepsy Foundation rare epilepsy network
epilepsy.com/what-is-epilepsy/rare-epilepsies/ren

Epilepsy Research Institute UK
epilepsy-institute.org.uk
Advocates and actions the research priorities of people affected by epilepsy.

European Epicare
epi-care.eu
The European Reference Network for all rare and complex epilepsies.

Trusted Information Creator
Patient Information Forum

Epilepsy Sparks
epilepsysparks.com/rare-epilepsy-organisations
List of international information/support groups.

Lennox Gastaut Syndrome
lgsfoundation.org
Support for Lennox Gastaut syndrome in the UK.

Rare epilepsy network
rareepilepsynetwork.org
Aims to improve outcomes for people with rare epilepsies. USA based.

Rett Syndrome
rettsyndrome.org.uk
Information on Rett Syndrome.

Ring Chromosome 20 Syndrome
ring20researchsupport.co.uk
Research, information and support for Ring Chromosome 20 Syndrome.

Sturge Weber Foundation
sturgeweber.org.uk

West Syndrome
wssg.org.uk

UK rare epilepsies together
ukret.co.uk
A UK rare epilepsies group with a network of patient advocacy groups.

Young Epilepsy
youngepilepsy.org.uk
Information and support with epilepsy syndromes.

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For a printed copy of this information contact our helpline.

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