What is a syndrome?

A syndrome is a group of signs and symptoms which, if they occur together, can suggest a particular condition.

Many children and young people will have a set of related symptoms which fit a particular pattern and this will determine which syndrome they have.

Knowing which syndrome will help the doctors to choose the appropriate antiepileptic drug (if needed) and also give a more accurate prognosis for the child/young person. It can also help to point parents in the right direction to get help and support.

Knowing the syndrome will be useful for understanding:

- whether the child/young person’s seizures are likely to be controlled
- which medication is likely to work best
- whether there are likely to be any other problems, for example with behaviour, learning or social functioning
- long term prognosis

Lennox Gastaut Syndrome

Lennox Gastaut Syndrome (LGS) is a rare, but severe, type of epilepsy that affects approximately 1-5% of children with paediatric epilepsy.

The age of onset ranges between 3-5 years old, with slightly more boys being affected than girls.

Although some children will have developmental delay prior to the onset of seizures, most children will develop normally prior to the onset of seizures and then begin to lose skills. The skills then tend to plateau.

The most common type of seizures experienced by children with LGS are atonic seizures. As children with this syndrome grow older they tend to develop a range of other seizure types.

Behavioural problems are common in LGS and may range from hyperactivity to autistic behaviours.

LGS has three defining characteristics:

- multiple seizure types
- a distinctive brain wave pattern
- learning difficulties which may range from slight to severe
Seizure types

The seizures that a child with LGS may experience may vary and include:

- **atonic seizures** (drop attacks) - these seizures cause a sudden loss of muscle tone resulting in the child dropping to the ground. They usually happen many times a day and often result in the child sustaining injuries that are so severe, protective headgear is required.

- **absence seizures** or staring episodes - the child stares blankly into space and becomes unresponsive for between a few seconds to a few minutes. There may be additional features such as head nodding or eye blinking.

- **tonic seizures** - the child experiences stiffening of the arms and legs and falls. There is no repetitive jerking. These seizures are the most common and occur especially during sleep. They range in duration from a few seconds to a few minutes. These seizures disturb the child’s sleep, but may not awaken the child and are often undetected because they are so brief.

- **tonic clonic seizures** - the child goes stiff and falls and has repetitive jerking.

- **focal seizures** - come from one area of the brain. What the seizure looks like depends on where in the brain it comes from and what function that area is responsible for.

- **myoclonic seizures** - the child has sudden muscle jerks.

These seizures typically occur very frequently, with some children having numerous episodes daily.

Children may also experience a loss of alertness that can last from several hours to weeks at a time. This is known as non convulsive status epilepticus and may require medical intervention to rectify.

Children with LGS may also develop behavioural and psychological issues which can include poor social skills and attention seeking behaviour. The causes of these may be difficult to establish as it may be due to ongoing electrical activity, side effects of medication or difficulty in understanding information.

The causes of Lennox Gastaut Syndrome

In up to a third of cases of LGS no cause can be found. For other children, causes may include any condition that produces major brain dysfunction.
Better futures for young lives with epilepsy

Examples include brain damage that occurs at or before birth (asphyxia, low birth weight and prematurity), infections of the brain (meningitis, encephalitis and rubella), genetic conditions such as tuberous sclerosis and in about a third of cases the child will have had a history of West syndrome (infantile spasms).

Diagnosing Lennox Gastaut Syndrome

The diagnosis of LGS may not be given very quickly. When taking the medical history the doctor will look for signs of prior seizures, especially infantile spasms.

They will also look for conditions such as mental regression, hearing and visual impairment and tuberous sclerosis that sometimes accompany the condition in children who have this syndrome.

An electroencephalogram (EEG) (a non-invasive painless test that records brain activity by picking up electrical signals given off by brain cells) will be helpful if done when awake and asleep, even when the child is not actually having any seizures.

The EEG will show a distinctive pattern whilst awake and in sleep. Flashing light and hyperventilation (rapid breathing) tests may also be carried out whilst the EEG is in progress.

Brain imaging will also be part of the evaluation because brain abnormalities are common with this syndrome.

Treating Lennox Gastaut syndrome

In many cases LGS proves resistant to antiepileptic medication, even when additional drugs in high doses are added to the treatment regime (polytherapy).

Treatment with medication aims for a balance between the best possible seizure control whilst avoiding potential toxic side effects such as drowsiness, fatigue, nausea and unsteadiness of movement.

If there is little or no improvement with medication, the ketogenic diet, vagus nerve stimulation, or a combination of these may be tried.

On occasions, and especially if a child is having multiple drop seizures causing injuries, an operation called a corpus callosotomy may be considered.

Antiepileptic drugs

The antiepileptic drugs most frequently used are sodium valproate (Epilim), lamotrigine (Lamictal), topiramate (Topamax), clobazam, phenytoin and rufinamide (Inovelon).

The ketogenic diet

Over one half of those children who are placed on this high fat, low carbohydrate and controlled protein diet have a 50% reduction of seizures or more. The diet requires strict supervision and even the slightest departure may cause the diet to lose its effect.
Vagus nerve stimulation
The vagus nerve stimulator (VNS) is a device similar to a pacemaker which is able to stimulate the left vagus nerve and is designed to help in the control of seizures.

The VNS is not effective for every individual with epilepsy but has shown to be effective in reducing seizures in approximately 50% of individuals with epilepsy.

Corpus callosotomy
The fibres that connect the two halves of the brain are known as the corpus callosum. These fibres are cut to prevent the seizure activity spreading throughout the brain. This is particularly helpful for drop attacks.

Prognosis
The prognosis for children with LGS can vary from child to child. The vast majority of children will continue to have seizures into adulthood and may never be able to live independently. They will need ongoing support with their learning and behavioural problems.

Further help and information

The LGS foundation
www.lgsfoundation.org/

Lennox Gastaut support group
Tel: 01664 454305

Tuberous Sclerosis Association
Tel: 0121 445 6970
www.tuberous-sclerosis.org

Contact a Family
Tel: 0808 808 3555
www.cafamily.org.uk

Vagus nerve stimulation
www.cyberonic.com

Ketogenic diet
www.matthewsfriends.org.uk