Better futures for young lives with epilepsy

Helpline: 01342 831342
Email:helpline@youngepilepsy.org.uk

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 that fit a particular pattern and this will determine which syndrome they have.

Identifying the 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

Landau Kleffner Syndrome

Landau Kleffner Syndrome (LKS) is a rare and under-diagnosed epilepsy syndrome. It usually occurs in children between the ages of 3 and 9 and affects the area of the brain that controls speech and comprehension.

These children will have developed speech normally and then, suddenly or gradually lose language skills for no apparent reason. LKS affects twice as many boys as girls. Along with the onset of seizures and language regression, behavioural changes may occur.

Seizures

Most children with LKS have recognisable seizures, which commonly occur during the night. The onset of these usually occurs before the initial language regression and can include:

focal seizures – these seizures come from one area of the brain; what the seizure looks like depends on the function of that area.

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prolonged absences – these seizures may occur many times a day and provoke a brief 'trance-like' state. The child will stare blankly into space and become unresponsive.

atonic seizures (also known as drop seizures) – during these seizures muscle tone is lost causing the child to flop and fall to the ground

Language regression

Whilst the child's language skills initially develop normally, children who develop LKS lose the ability to understand language and the ability to use speech.

This language regression can occur suddenly or may be of gradual onset. Initially many children are thought to have developed a hearing deficit because it appears that they have difficulty in distinguishing between various sounds.

Behavioural issues

Children with LKS have a tendency to develop behavioural issues such as: attention deficit hyperactivity disorder (ADHD), including over-activity, reduced concentration span, irritability, tantrums and difficulties with social interaction.

Diagnosis

The diagnosis of LKS is made on the basis of the child's history. Relevant features will be a history of normal early development followed by loss of language skills, often in association with seizures and changes in behaviour.

- **Electroencephalogram** (EEG) – this is a non-invasive, painless test that records brain activity by picking up the signals given off by brain cells. In LKS, recordings can prove helpful, especially in the active phase of the disorder.

  The EEG will usually show abnormalities often concentrated in the area of the centro-temporal regions, which are known to be important for language.

  These abnormalities are more obvious during sleep and some of the activity results in actual seizures during the active phase of the condition. However much of the activity is 'sub-clinical', and will present as continuous spike-and-wave (CSWS) discharges in sleep.

  For this reason a sleep record is usually required and may involve the child undergoing video telemetry. This is a type of EEG that involves day and night recording with simultaneous videoing of the child.

- **Scans** – MRI and CT scans are usually normal with no abnormalities visible.

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Treatment

Management of LKS involves treating the seizure activity to reduce the effect on the child and providing the necessary support to ensure effective recovery.

Treating the seizures
The drugs commonly used to treat the seizures are sodium valproate (Epilim), ethosuximide (Zarontin) and benzodiazepines such as clobazam (Frisium). Steroids may also be prescribed, often given in weekly pulses.

Speech and language therapy
Speech and language therapy is a vital part of the management of LKS and assessing and monitoring progress is an important part of dealing with this syndrome.

Brain surgery
Previously an operation called multiple subpial transaction was performed, but is not used much nowadays due to lack of consistent efficiency.

Prognosis

During the active phase of LKS there may be repeated episodes of regression and recovery, and a child's abilities and performance may be extremely variable. Seizures are usually controlled with antiepileptic drugs and the EEG abnormalities may disappear.

Although some children will make a good recovery, many children are left with significant impairments. The overall outcome is thought to be related to the length of time of the active phase of LKS.

Generally it is thought that about 50% of children will make a reasonable recovery, 25% will have a partial recovery and a further 25% have very significant persisting difficulties with language, behaviour or cognitive skills.

For more information and support

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

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