

# Glossary

## Landau-Kleffner Syndrome (LKS)

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### **What is Landau-Kleffner Syndrome?**

Landau-Kleffner Syndrome (LKS) is a rare type of childhood epilepsy syndrome that occurs in approximately one child in a million, in children usually between the ages of three and nine years. LKS is more common in boys and does not usually run in families. The underlying cause(s) of LKS is not yet known.

The main features of LKS are a loss of understanding and speaking words, with epileptic seizures.

### **Features of Landau-Kleffner Syndrome**

#### **i) Epileptic seizures**

Epileptic seizures develop in about three quarters of children with LKS. Epileptic seizures happen when part of the brain develops uncontrolled electrical activity, which stops the normal function of that part of the brain.

Epileptic seizures in LKS usually involve the type of seizures that occur silently during sleep. However, there may also be other types of seizures.

#### **ii) Speech and language skills**

In LKS the child generally loses understanding first, then speech. The difficulty with understanding may become obvious if the child is not able to recognise voices, or the content of conversations, or tell the difference between everyday sounds. This loss may be sudden (even overnight), or gradual over a period of months and is often mistaken for deafness initially. Many children compensate by using visual cues, gestures and by copying others and may initially hide their difficulty. Children may have difficulties expressing themselves, may talk nonsense or in jargon, or even stop speaking altogether. The deterioration in skills is called a regression, as the child appears to have returned to an earlier stage in their development.

#### **iii) Additional difficulties**

LKS was initially thought to be specific to language, but it is now clear that other abilities are also often affected.

Behavioural changes are commonly reported and may include over-activity, reduced concentration span, irritability, tantrums and difficulties with social interaction. The behavioural changes often prevent children from learning and socialising well. The severity of the language and behavioural problems varies over time. Non-verbal thinking (cognitive) skills are usually relatively unaffected, although there may be isolated difficulty, for example with thinking speed.

The child may also have problems with fine motor coordination and movement, such as dribbling, messy eating, loss of speech clarity, clumsiness and shakiness. These difficulties are thought to be a direct result of the disease process, rather than simply an emotional reaction to loss of language.

### **Assessment and diagnosis**

As LKS is so rare, it is common for children to be investigated for deafness, autism, selective mutism, verbal dyspraxia or behavioural problems before the true diagnosis of LKS is made.

The diagnosis of LKS is made on the basis of a medical interview and examination. The core features are a history of normal early language development followed by loss of language skills, often in association with mild seizures and behavioural changes. Physical examination is usually normal, except for occasional difficulty with movement or coordination. Brain imaging is usually normal. Electroencephalograms (EEG) of brainwave activity in LKS show abnormal electrical changes over the language areas of the brain and these often increase in frequency during sleep.

## Treatment and management

Assessment by a multidisciplinary team including medical, speech and language therapy and clinical psychology services, is an integral part of management. It enables a holistic approach to assess and monitor response to treatment, as well as identifying appropriate therapy and educational interventions. Repeat sleep EEG records are also used to monitor treatment.

Treatment is usually aimed at the electrical seizures that occur in sleep and typically involves medication. Continued monitoring of a child's understanding and talking is also important to monitor the effectiveness of treatments, as there is not always a straightforward relationship between abnormalities detected by an EEG and the child's language skills.

## Progress

LKS is not usually life threatening, but can impact greatly on quality of life unless it responds well to treatment. The course of LKS is variable and speech and language skills may improve over time.

The active phase can last some years, but typically 'burns out' by early adolescence. During the active phase, the child will be vulnerable to further regression and fluctuation but usually the first regression is the most severe and in many children, medication can help to recover skills and prevent further relapse.

Children may lose skills and then regain them. There is often extreme fluctuation in how a child can access their skills. Therefore a child's abilities may appear to change dramatically, for better or worse, over short periods of time, even within a day. This lack of predictability and extreme fluctuation makes adjustment for families and school very difficult.

The best treatment response appears to be seen in children whose regression is largely limited to the ability to understand speech, without additional difficulty with behaviour, social communication and general learning. It is also important that treatment is given early.

Once the active phase is over, there is often a period of natural recovery. The extreme fluctuation settles and the child's good skills and remaining areas of difficulty become clearer.

The recovery phase may last well into early adulthood and it is important to take this into account in relation to ongoing educational support.

## Support

LKS is a rare diagnosis and there is often little knowledge amongst local professionals about the syndrome. This means that parents/carers may have to invest a significant amount of time speaking to local education and health services about their child's often fluctuating needs and the importance of timely support. A number of organisations can offer information and support.

## References

### Epilepsy Action

<https://www.epilepsy.org.uk/info/syndromes/landau-kleffner-syndrome>

### Young Epilepsy

<http://www.youngepilepsy.org.uk/dmdocuments/Landau-Kleffner.pdf>

### Great Ormond Street Hospital

<https://www.gosh.nhs.uk/medical-information/search-medical-conditions/landau-kleffner-syndrome>

### Contact

<http://www.contact.org.uk/medical-information/conditions/l/landau-kleffner-syndrome/>

**Please note: Afasic does not hold copies of any referenced material. These publications should be available at academic libraries.**

## Organisations which can help

### Cerebra

2nd Floor Offices  
The Lyric Buildings  
King Street  
Carmarthen  
SA31 1BD

<http://w3.cerebra.org.uk/>

Tel: 01267 244200

Helpline Freephone: 0800 328 1159

E mail: [enquiries@cerebra.org.uk](mailto:enquiries@cerebra.org.uk)

Working for children with brain conditions

### Contact

209-211 City Road  
London  
EC1V 1JN

<http://www.contact.org.uk>

Tel: 020 7608 8700

E mail: [info@contact.org.uk](mailto:info@contact.org.uk)

**Epilepsy Action**

New Anstey House  
Gate Way Drive  
Yeadon  
Leeds  
LS19 7XY

<https://www.epilepsy.org.uk>

Helpline: 0808 800 5050 (international +44 113 210 8800)

Email: [epilepsy@epilepsy.org.uk](mailto:epilepsy@epilepsy.org.uk)

You can also find them on Facebook, Twitter, and YouTube

**Great Ormond Street Hospital**

Wolfson Neurodisability Service  
Level 10  
Main Nurses Home  
Great Ormond Street Hospital  
Great Ormond Street  
London  
WC1N 3JH

<http://www.gosh.nhs.uk/parents-and-visitors/clinical-support-services/occupational-therapy/services-we-provide/neurodisability-services>

Tel: Developmental Epilepsy Clinic  
020 7405 9200, ext 1144

**Royal College of Speech and Language Therapists (RCSLT)**

2 White Hart Yard  
London  
SE1 1NX

<https://www.rcslt.org/>

Tel: 020 7378 1200

The professional body for speech and language therapists

**Young Epilepsy**

St. Piers Lane  
Lingfield  
Surrey  
RH7 6PW

<http://www.youngepilepsy.org.uk>

Tel: 01342 832243 (main switchboard)

Helpline: 01342 831342

Email: [info@youngepilepsy.org.uk](mailto:info@youngepilepsy.org.uk)

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