

Landau-Kleffner syndrome is a rare condition in which children lose the ability to speak and to understand speech. This change is associated with seizures, which are usually infrequent or easily controlled with medication, and with a specific EEG pattern known as electrical status epilepticus in sleep (ESES).

Landau-Kleffner syndrome seems to be part of a continuum of syndromes that involve an abnormal EEG pattern and various different symptoms. A related syndrome is epilepsy with continuous spikes and waves during slow sleep (CSWS), which is also discussed on this page.

What are other terms for Landau-Kleffner syndrome?

Other terms for Landau-Kleffner syndrome that you may come across include:

- acquired childhood aphasia with convulsive disorder
- acquired epileptic aphasia
- Landau-Kleffner acquired epileptiform aphasia

What causes Landau-Kleffner syndrome?

The cause is not known. In some children, a focal brain lesion may produce the abnormal EEG patterns and seizures that are seen with Landau-Kleffner syndrome.

Landau-Kleffner syndrome may be caused by any underlying brain problem, including:

- congenital malformation
- cyst

- brain tumour

Rarely, BECTS may evolve into Landau-Kleffner syndrome.

What are the features of Landau-Kleffner syndrome?

Landau-Kleffner syndrome usually begins when the child is between three and eight years old. A child who has learnt to speak gradually loses the ability to understand speech, and stops responding to spoken words. The child's parents and doctor may assume at first that he has lost his hearing.

The child may begin to stutter or to make mistakes in his speech, such as mispronouncing words or using the wrong words or the wrong combinations of words. He talks less and less. In severe cases, he may stop talking altogether, and may not even respond to non-verbal sounds such as a ringing telephone.

In some cases, children with Landau-Kleffner syndrome can still communicate without speech, for instance by writing or using sign language.

The child may develop behaviour problems, including hyperactivity, excitability, and attention deficit. In some cases, the child's mental and behaviour problems become so severe that he cannot function normally.

Children with Landau-Kleffner syndrome have a characteristic EEG pattern known as electrical status epilepticus in sleep (ESES). During sleep, the EEG pattern is dominated by spike and wave discharges. These abnormalities may continue

when the child is awake. Children with Landau-Kleffner syndrome also have seizures. These may be partial motor seizures, generalized tonic-clonic seizures, atypical absence seizures, eye blinking, or atonic seizures.

Typically, the seizures are less severe than the child's language problems. Seizures are usually easily controlled with anti-epileptic medications. The seizures and the abnormal EEG patterns usually go away by the time the child reaches age 15, and the language problems may improve somewhat at this point.

How many other children have Landau-Kleffner syndrome?

We do not know how many children have Landau-Kleffner syndrome, but it is quite rare.

How do you know that a child has Landau-Kleffner syndrome?

Landau-Kleffner syndrome may be confused with deafness, pervasive developmental delay, or autism. To diagnose Landau-Kleffner syndrome, it is important for the child's doctor to understand his development, particularly in terms of language. His doctor may ask to see school reports and any other reports on his behaviour and development. She may order or perform a variety of tests, such as:

- neuropsychological testing to evaluate how his brain processes language and performs other functions, and to find out the specific type of language problem that is affecting him
- EEG while the child is awake and asleep
- in rare cases, video-EEG
- neuroimaging (CT or MRI)

How is Landau-Kleffner syndrome treated?

Because Landau-Kleffner syndrome is so rare and its outlook varies, there have been no controlled trials to study its treatment. What we know about the treatment options for this syndrome comes only from case reports.

The seizures caused by Landau-Kleffner syndrome are usually mild and easy to control. Most anti-epileptic drugs are effective in preventing the seizures, but have little effect on behaviour or language problems. For the language difficulties, high-dose corticosteroids seem to give the best results. Benzodiazepines may also be tried. Intravenous immune globulin (IVIG) also appears to have good results in some cases.

Two surgical procedures, multiple subpial transection and vagal nerve stimulation (VNS), have been considered, although the studies that looked at them were very small. In rare conditions such as Landau-Kleffner syndrome, treatment must be carefully tailored to each child by a physician experienced in the care of children with the condition.

Most importantly, children with Landau-Kleffner syndrome may need special teaching that takes their language problems into account. Each child has his own particular needs, but some strategies that may be helpful include:

- a helper in the classroom to give the child one-on-one help with understanding and presenting information

- using sign language, pictures, colour coding, and other visual cues to help the child understand spoken information
- computer programs with colourful images and very simple verbal information
- speech and language therapy

Teachers need to be educated about the child's condition so that they can give him the support he needs and understand that the child may have problems processing spoken language for some time.

More information

- Drug Therapies for Epilepsy
- Surgical Treatment of Epilepsy

What is the outlook for a child with Landau-Kleffner syndrome?

The outlook for children with Landau-Kleffner syndrome varies. The seizures can usually be controlled with anti-epileptic drugs, and the seizures and abnormal EEG patterns usually stop by themselves after a few years. Fewer than 20% of children with this syndrome continue to have seizures, and they are usually rare.

Once the EEG abnormalities improve, the child's language abilities may also improve and he may be better able to learn. However, the child will usually need intensive language therapy and help with school, even after the seizures are under control.

In at least 50% of cases, children may have permanent problems with language and verbal short-term memory. These may affect a child's education and his everyday life, even as an

adult. The outlook for his language abilities depends on:

- how old the child was when the syndrome began; the younger he was, the more severe the problems, because the syndrome was active while he was developing language skills
- how severe the EEG disturbances were
- the location of the disturbances
- how long the EEG disturbances continued

There is no connection between the number of seizures a child has and the outcome of his language abilities.

Epilepsy with continuous spikes and waves during slow sleep (CSWS)

This syndrome is related to Landau-Kleffner syndrome, in that they share the same EEG pattern, known as electrical status epilepticus during slow sleep (ESES). Epilepsy with CSWS is also fairly rare. It can result in memory, language, and behaviour problems and lowered IQ. The cause of the syndrome is unknown. It does not seem to run in families.

The syndrome affects more boys than girls. It begins between the ages of one and 11 years, with most children having their first seizure at about four to six years of age. Children with this syndrome may have developed normally before the seizures began, or may have had abnormal neurological findings such as one-sided weakness or paralysis or movement problems.

The syndrome may first appear as generalized clonic seizures, often at night. At the same time or later, the EEG abnormality of continuous

spikes and waves during slow sleep (CSWS) appears. The seizures become more severe, and most children will have two or three different seizure types. Atypical absence seizures are the most common, but partial motor seizures, complex partial seizures, generalized clonic seizures, and tonic-clonic seizures are also seen. The seizures may happen while the child is awake or asleep. Absence status epilepticus is seen in about half of affected children.

At the same time, the child's mental development slows down, stops, or even goes backward. The child usually has problems with language and may also have problems with memory, movement, and behaviour, including loss of coordination, involuntary movements or spasms, aggressiveness, or hyperactivity.

The seizures and CSWS normally stop before or near puberty; on average, the epilepsy lasts for 12 years. There is a slow improvement in the child's movement abilities. However, the mental and behavioural problems often continue.

The syndrome may be confused with Lennox-Gastaut syndrome, Landau-Kleffner syndrome, or BECTS. CSWS is distinguished from these syndromes primarily by the neuropsychological and motor regression seen in CSWS. To diagnose the syndrome, the child's doctor may order an all-night EEG recording during sleep.

The seizures are treated according to the type of seizure. They can usually be controlled with anti-epileptic drugs. There is no specific treatment that is always helpful in treating the other problems associated with this syndrome. Corticosteroids may produce some improvement with the mental problems. As with all rare conditions, treatment must be carefully tailored to each child by a physician experienced in the care of children with the condition.

About half of children with this syndrome never return to normal levels of mental and behavioural functioning, especially in language and attention. The outcome may be related to the length of time the CSWS lasted.