Landau-Kleffner syndrome

What is Landau-Kleffner syndrome?
Landau-Kleffner syndrome (LKS) is a rare form of epilepsy and usually begins in children between 2 and 8 years old. The most common average age of onset is 5 to 7 years of age and it affects both sexes equally. A progressive loss of speech is typically seen in a child with previous age-appropriate development. The language disorder may start suddenly or slowly. It usually affects the child's understanding of spoken language the most. Yet it may affect both understanding speech and speaking ability, or it may affect speaking only. Attention deficit problems – with or without hyperactivity, anxiety, and aggression – can be seen in almost 8 out of 10 children.

What are the symptoms of Landau-Kleffner syndrome?
Seizures occur in 3 out of 4 children with this epilepsy syndrome.

- The most common seizure type seen in LKS is focal motor seizure.
- Focal seizures can become tonic-clonic seizures, also called bilateral convulsion.
- Atypical absence and atonic seizures have been reported.
- Seizures typically occur in sleep and are not very frequent.

The cause of LKS is unknown, though new (de novo) genetic mutations have been found in children. A gene called GRIN2A is known to be involved. A family history of seizures or epilepsy is not typically seen.

How is Landau-Kleffner syndrome Disorder diagnosed?

- The first step is getting a good history and examination of your child.
- An EEG (electroencephalogram), and in many instances a prolonged EEG or video EEG, is needed to diagnose LKS. The EEG in children with this epilepsy syndrome is abnormal, especially as they enter sleep. Frequent high amplitude spike and sharp waves in the temporal and parietal lobes are seen primarily during sleep.
- Genetic and metabolic tests may be ordered by your doctor.
- An MRI (magnetic resonance imaging) scan is typically normal.

Treatment options for Landau-Kleffner syndrome

- Seizures in children with LKS can be treated with anti-seizure medications. In rare instances, surgery may be done.
- Medications often used are steroids, valproic acid (Depakote/Depakene) or high dose diazepam. Other medications that have been used to treat this epilepsy syndrome include clobazam (Onfi), ethosuximide (Zarontin), topiramate (Topamax), and levetiracetam (Keppra). Often, more than one medicine is used at once.
- Sometimes seizures persist despite medication. In those children, epilepsy surgery at an epilepsy center may become an option. A type of surgery called multiple subpial transections in which multiple small cuts are made in the brain cortex may be possible.
- Resective brain surgery is not usually an option.
- Speech therapy should be started immediately in children with LKS.
- Children may need to see child psychologist, neuropsychologist, and/or psychiatrist to treat symptoms of ADHD (attention deficit hyperactivity disorder), anxiety, or aggression.
- Children with LKS rarely have cluster seizures. If they do happen, emergency medical treatment or treatment with a rescue therapy may be needed, for example diazepam rectal gel (Diastat) or another form on benzodiazepine given into the nose (intranasally) or under the tongue.
- Parents of children with Landau-Kleffner syndrome should talk to the treating neurologist or health care provider to learn about seizure emergencies.
- Talk to the health care team about what kind of rescue therapy could be used and when to use it.
- When seizures last longer than usual or if a generalized seizure lasts too long (generally considered 5 minutes or longer), a child may need emergency medical care.

Long-term complications of Landau-Kleffner syndrome

- The outlook for recovering language function is poor when LKS starts early in life.
- Seizures and EEG abnormalities go away in most cases. After age 10, only 1 or 2 out of 10 children with LKS will still have seizures. The course of the disorder changes, and it occasionally disappears on its own.
- Over 8 of 10 children are left with permanent language difficulties and in many instances these can be severe. This is most commonly seen in children when medical or surgical therapy does not eliminate the epileptic patterns on the EEG.

Resources

- **Landau-Kleffner Syndrome: Epilepsy Foundation -** www.epilepsy.com/learn/types-epilepsy-syndromes/landau-kleffner-syndrome
- **National Organization for Rare Disorders** - https://rarediseases.org/rare-diseases/landau-kleffner-syndrome