Lennox-Gestaut Syndrome

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Lennox-Gestaut syndrome (LGS) is a form of early childhood-onset epilepsy that most often appears between the second and sixth years of age though this is not exclusive. It can appear at any time although it frequently occurs in children with a prior history of epilepsy and evidence indicates that LGS closely parallels West Syndrome, which is another type of epilepsy. LGS is characterized by frequent seizures of different types giving this form of epilepsy the broadest range of seizure types than any other type.

- Commonly occurring seizure types
    - Tonic, of which ninety percent are nocturnal
    - Myoclonic (muscle spaticity)
    - Mixture of atonic, atypical absence (petit mal), complex partial, focalized, and generalized tonic-clonic (grand mal) seizures.
- Affects males more often than females and approximately five percent of all children having epilepsy have Lennox-Gestaut. These children frequently have halted or slowed psychomotor development.
- Behavior disorders are common.
- There is no uniform cause for LGS and eighty percent of cases are idiopathic, meaning they do not have an identifiable underlying cause.
- Twenty percent of cases have identifiable causes that include but are not limited to encephalopathy, tuberous sclerosis, metabolic diseases, encephalitis, meningitis, toxoplasmosis, hypoxic-ischemic injury, and frontal lobe lesions.
- First-line drugs include valproic acid, sodium valproate, felbamate, and benzodiazepines, specifically clonazepam.
- Second-line drugs used for treatment are topiramates and lamotrigine.
- Surgical options may include insertion of a vagal nerve stimulator to potentially reduce the number of seizures or performing a corpus callostomy, an operation that severs the corpus callosum , which is a band of nerve tissue connecting the two hemispheres of the brain.
- Other treatments suggest the use of a ketogenic diet, which is gaining popularity although there is minimal mainstream acceptance.
- Immunoglobulin therapy can help reduce seizure frequency and length in some cases.
- There is no cure for LGS but drug therapy can provide a higher quality of life and functionality for these patients.