



Lennox-Gastaut Syndrome

Epilepsy Action is indebted to Dr Richard Appleton, a consultant who specialises in children's epilepsy, and his associates, at Alder Hey Hospital, Liverpool, who have kindly written this factsheet.

For further information about epilepsy or anything mentioned in this factsheet, please contact the Epilepsy Helpline freephone 0808 800 5050 or helpline@epilepsy.org.uk.

What is a syndrome?

A syndrome is a group of signs and symptoms that, added together, suggest a particular medical condition. In epilepsy, examples of these signs and symptoms would be things like the age at which seizures begin, the type of seizures, whether the child is male or female and whether they experience difficulties with learning.

Lennox-Gastaut Syndrome

This type of epilepsy is uncommon and occurs in between one and five in every 100 children with epilepsy. However, it is the most common cause of intractable (difficult to treat) childhood epilepsy. The most common time for it to start is between three and five years of age.

Symptoms

The seizures experienced can vary. The most common type is atonic seizures or 'drop attacks'. During these, the child will suddenly fall to the ground which may at first be mistaken as tripping up or poor balance. These seizures usually happen

many times a day and are very upsetting as the child is often injured during them. Another common seizure type is atypical absence seizures, which can last from 10 seconds to several minutes. During these seizures the child will appear vacant or blank. They may have other features like head nodding, or rapid blinking. The child may have some awareness of what is going on around them. These seizures can be very frequent and can even merge to be constant, forming a state called 'non-convulsive status epilepticus'. During these episodes the child will 'not be with it', drool, be unable or slow to speak, need help with feeding and be floppy or wobbly.

Another frequent seizure type is tonic seizures. They may only happen at night. There is a general stiffening of the arms and legs. Children with Lennox-Gastaut syndrome may have other types of seizures including generalised tonic-clonic seizures, partial motor seizures, complex partial seizures and myoclonic seizures.

All children with Lennox-Gastaut syndrome will develop learning difficulties which will be moderate to severe. Some children will have developmental delay or learning difficulties before the seizures start. About two out of ten children will develop Lennox-Gastaut syndrome after having West syndrome* (infantile spasms).

Diagnosis

The diagnosis becomes clearer with time. The electroencephalogram (EEG) is very helpful as typical abnormalities are seen, even when the child

is not having any obvious seizures. Some children may have a cause which can be identified. These included a genetic condition called *tuberous sclerosis, a structural brain malformation, brain damage due to problems before or around the time of birth, meningitis as a young baby or a head injury. In about half the cases no cause is found.

Treatment

Treatment of Lennox-Gastaut syndrome is very difficult. The drugs which may be effective in some cases include sodium valproate, lamotrigine, topiramate, clobazam and phenytoin. Sometimes the ketogenic diet* may be effective. Courses of corticosteroid drugs are sometimes used when seizures are particularly difficult to control.

In children who have repeated drop attacks surgery may be helpful. Corpus callostomy* and Vagus Nerve Stimulation (VNS) * are two surgical procedures that may be considered.

Prognosis (outlook)

The long term prognosis is poor in terms of seizure control and intellectual development. A very small number of children will outgrow their seizures by their teenage years. However, the remaining children will continue to have seizures, often on a daily basis, into adult life. These patients will be dependent on others for all their life. Some children will develop problems like repeated or severe chest infections which may shorten their life.

* Factsheets on West syndrome, ketogenic diet and epilepsy surgery are available from Epilepsy Action.

For information on seizure types, anti-epileptic medication and general aspects of epilepsy please contact the Epilepsy Helpline freephone 0808 800 5050.

Support organisation

Lennox-Gastaut Support Group, 9 South View, Burrough on the Hill, Melton Mowbray, LE14 2JJ, telephone 01664 454 305.

Tuberous Sclerosis Association, P O Box 12979, Barnt Green, Birmingham, B45 5AN, telephone 0121 445 6970, www.tuberoussclerosis.org.uk

Contact a Family, 209-211 City Road, London, EC1V 1JN, telephone 0808 808 3555, www.cafamily.org.uk

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