Landau Kleffner Syndrome (LKS)

What is Landau Kleffner Syndrome (LKS)?
Landau Kleffner Syndrome is a rare form of childhood epilepsy which results in a severe language disorder. It is also known as ‘acquired epileptic aphasia’. LKS affects girls and boys almost equally.

The disorder usually starts between 4 and 7 years of age, though it may occur in children as young as 2 or as old as 11. Typically the first indication that something is wrong is that the child’s understanding of language deteriorates. In most speech and language disorders in children the child’s language will always have lagged behind; but in LKS there is a loss of language skills in a child who was previously developing normally and who was beyond the ‘first words’ stage. This deterioration of language can happen slowly, over a period of weeks or months, or much more quickly, over days.

The language disorder in most children affects understanding of spoken language; the ability to speak is usually seriously affected too. The child may not respond to environmental sounds, like a doorbell, phone or vacuum cleaner.

Children with LKS are often thought to be deaf because of their difficulty in understanding what people say to them. However, hearing tests show that the child can detect sounds: the difficulty lies in interpretation of the sounds.

In some cases a diagnosis of selective mutism may be suspected. Unlike the child with selective mutism, the child with LKS will fail to speak in all situations, not just in some. Furthermore, children with selective mutism do understand what is said to them, whereas this is typically not the case in LKS.

Sometimes it may be difficult to distinguish between LKS and autistic disorder. Children with autism often have severe difficulties understanding spoken language, and they too may lose language skills after a period of normal development. But in autism difficulties begin earlier (usually being apparent to parents by 30-36 months of age) and the child has difficulties with all aspects of social interaction and communication. Children with LKS, by contrast, will respond socially in a normal way, and will communicate in ways other than spoken language – for instance, through gestures and facial expressions.

This does not mean that the difficulties for a child with LKS are restricted to language. Some children are frustrated or frightened by their inability to communicate: this can cause temper tantrums, withdrawal from the world or other behavioural difficulties. If the epileptic activity affects the frontal regions of the brain, children may be uninhibited and lack restraint in their behaviour.

LKS can be difficult to diagnose because many children do not have obvious epileptic seizures. Abnormal epileptic activity, in one or both temporal lobes of the brain, does show up on a test called an EEG (electroencephalogram). The temporal lobes are important for language comprehension and memory. There is no evidence that the brain is damaged in LKS; but the underlying epileptic activity interferes with the child’s ability to learn and understand language.

In some children the communication disorder fluctuates, so that speech and language may improve and then get worse again. Older children, especially those aged 6 and above when the disease starts, will often make a good recovery. For younger children (those less than 4 years at onset) the ability to understand speech may remain seriously impaired – although some younger children do make a good recovery after an initial period of deterioration. As for seizures, the outcome is usually good, and the EEG becomes normal.

As LKS is rare, many paediatricians will never have encountered a case; thus the reason for the child’s communication difficulties can go unrecognised. Any child who develops serious communication difficulties after a period of normal development should be seen by a specialist with expertise in epilepsy, who will investigate any underlying epilepsy and recommend the appropriate treatment.
Medical treatment
Medical treatments are not usually very effective, but it can be of benefit to control the epileptic activity at an early stage. Drugs called corticosteroids can help – but these are powerful and most paediatricians will use them very cautiously. Other anti-epileptic medicines can control epileptic activity, but do not always improve language. Some children have benefited from a specific method of brain surgery – but this is not appropriate for all children with LKS.

Educational implications
Most children benefit from alternative communication methods, especially sign language. Those with persistent language impairment usually need special education with other language-impaired children in a school where sign language is used by teachers and other pupils. Children with LKS are sometimes educated with children who have hearing impairments. Parents of children with LKS are advised to learn sign language, so that they can communicate more easily with their child.

References

Other relevant Glossary Sheets
- Selective mutism (6)
- Epilepsy and speech and language (24)

Other organisations which can help
Epilepsy Action
New Anstey House
Gate Way Drive
Yeadon
Leeds
LS3 1BE
Helpline: 0800 800 5050

F.O.L.K.S. (Friends of Landau-Kleffner Syndrome)
Ground Floor
3 Stone Buildings
Lincolns Inn
London
WC2A 3XL
Helpline: 0870 847 0707

National Society for Epilepsy
Chesham Lane
Chalfont St Peter
Bucks
SL9 0RJ
Tel: 01494 601300

Written by Dr Gillian Baird, Newcomen Centre, Guy’s Hospital, and Dr Dorothy Bishop, MRC Applied Psychology Unit, Cambridge

© Afasic 2004

Afasic
2nd Floor
50-52 Great Sutton Street
London EC1V 0DJ
Phone 020 7490 9410
Fax 020 7251 2834
Email info@afasic.org.uk
www.afasic.org.uk
Helpline 0845 3 55 55 77
(local call rate)

Registered charity no. 1045617