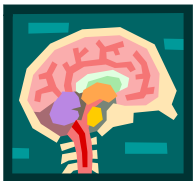

Disability Awareness Begins With You:

Landau-Kleffner Syndrome

What is Landau-Kleffner syndrome?

Landau-Kleffner syndrome (LKS) is a childhood disorder. A major feature of LKS is the gradual or sudden loss of the ability to understand and use spoken language. All children with LKS have abnormal electrical brain waves that can be documented by an electroencephalogram (EEG), a recording of the electric activity of the brain. Approximately 80 percent of the children with LKS have one or more epileptic seizures that usually occur at night. Behavioral disorders such as hyperactivity, aggressiveness and depression can also accompany this disorder. LKS may also be called infantile acquired aphasia, acquired epileptic aphasia or aphasia with convulsive disorder. This syndrome was first described in 1957 by Dr. William M. Landau and Dr. Frank R. Kleffner, who identified six children with the disorder.

What are the signs of Landau-Kleffner syndrome?

LKS occurs most frequently in normally developing children who are between 3 and 7 years of age. For no apparent reason, these children begin having trouble understanding what is said to them. Doctors often refer to this problem as auditory agnosiaor "word deafness." The auditory agnosia may occur slowly or very quickly. Parents often think that the child is developing a hearing problem or has become suddenly deaf. Hearing tests, however, show normal hearing. Children may also appear to be autistic or developmentally delayed.

The inability to understand language eventually affects the child's spoken language which may progress to a complete loss of the ability to speak (mutism). Children who have learned to read and write before the onset of auditory agnosia can often continue communicating through written language. Some children develop a type of gestural communication or sign-like language. The communication problems may lead to behavioral or psychological problems. Intelligence usually appears to be unaffected.

The loss of language may be preceded by an epileptic seizure that usually occurs at night. At some time, 80 percent of children with LKS have one or more seizures. The seizures usually stop by the time the child becomes a teenager. All LKS children have abnormal electrical brain activity on both the right and left sides of their brains.

How common is Landau-Kleffner syndrome?

More than 160 cases have been reported from 1957 through 1990.

What causes Landau-Kleffner syndrome?

The cause of LKS is unknown. Some experts think there is more than one cause for this disorder. All of the children with LKS appear to be perfectly normal until their first seizure or the start of language problems. There have been no reports of children who have a family history of LKS. Therefore, LKS is not likely to be an inherited disorder.

What is the outcome of Landau-Kleffner syndrome?

There have not been many long-term follow-up studies of children with LKS. This lack of evidence, along with the wide range of differences among affected children, makes it impossible to predict the outcome of this disorder. Complete language recovery has been reported; however, language problems usually continue into adulthood. The continued language problems can range from difficulty following simple commands to no verbal communication. If recovery takes place, it can occur within days or years. So far, no relationship has been found between the extent of the language impairment, the presence or absence of seizures and the amount of language recovery. Generally, the earlier the disorder begins, the poorer the language recovery.

Most children outgrow the seizures, and electrical brain activity on the EEG usually returns to normal by age 15.

What treatments are available?

Medication to control the seizures and abnormal brain wave activity (anticonvulsants) usually has very little effect on language ability. Corticosteroid therapy has improved the language ability of some children. Sign language instruction has benefited others.

Where can I get more information?

NIDCD maintains a directory of organizations that can answer questions and provide printed or electronic information on Landau-Kleffner syndrome. Please see the list of organizations at www.nidcd.nih.gov/directory.

For more information, additional addresses and phone numbers, or a printed list of organizations, contact:

NIDCD Information Clearinghouse
1 Communication Avenue
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http://www.nidcd.nih.gov/health/voice/landklfs.htm