

## **Managing Landau - Kleffner Syndrome**

Landau-Kleffner syndrome (LKS) is an acquired epileptic aphasia of childhood and is a rare, childhood neurological syndrome. It is accompanied by abnormal electroencephalogram (EEG) and behaviour symptoms of autism.

LKS may also be called infantile 'acquired aphasia', 'acquired epileptic aphasia' or 'acquired aphasia with convulsive disorder', but Rapin et al (1977) called it 'auditory verbal agnosia'. It was first described by Landau and Kleffner, who identified six children with LKS (1957).

Auditory verbal agnosia or 'pure word deafness' is involved — in other words, a disturbance in comprehension of spoken language in the presence of otherwise intact auditory functioning and, essentially, normal performance in other language modalities.

Aphasia itself is a defect or loss of the power of expression by speech, writing or signs, or a defect or loss of the power of comprehension of spoken or written language (the Greek word for 'speechless' is aphatos).

In terms of incidence, LKS usually starts before the age of six years and affects twice as many boys as girls (Beaumanoir et al, 1985). More than 198 cases have been reported from 1957 to 1992 (Beaumanoir, 1992).

The ICD 10 defines acquired aphasia with epilepsy/Landau-Kleffner syndrome as a "disorder in which the child, having previously made normal progress in language development, loses both receptive and expressive language skills but retains general intelligence. The onset of the disorder is accompanied by paroxysmal abnormalities on the EEG and, in the majority of cases, also by epileptic seizures".

### **Onset of seizures**

Usually, the onset is between the ages of three and seven years, with skills being lost over days or weeks. The temporal association between the onset of seizures and loss of language is variable, with one preceding the other (either way round) by a few months to two years. An inflammatory encephalitic process has been suggested as a possible cause of this disorder.

About two-thirds of patients are left with a more or less severe receptive language deficit. Physicians are advised to exclude aphasia due to causes not otherwise specified, autism or Heller's syndrome.

The exact cause of LKS is unknown. However, it is unlikely to be an inherited disorder, but overt cerebral pathology such as arteritis, tumours and inflammation may be associated with it (Smith et al 1992, De Volder et al, 1994). Toxoplasmosis (Maichalowicz et al, 1989), neurocystercosis (Otero et al, 1989), temporal astrocytoma (Solomon et al, 1993), haemophilus influenza meningitis (Aninsk et al, 1989), subacute sclerosing panencephalitis (Bicknese et al, 1996),

inflammatory demyelinating disease (Fayad et al, 1997) and zinc metabolism (Lerman-Sagie et al, 1987) are also all implicated in LKS.

Landau-Kleffner syndrome shares a common pathological mechanism with continuous spike-wave of sleep (CSWS), acquired epileptic opercular syndrome (AEOS) and benign childhood epilepsy with centro-temporal spikes (BECTS).

### **Characteristics**

The main feature of LKS is the gradual or sudden loss of the ability to understand and use spoken language. The speech and language skills may improve over time. Patients typically have:

- ⤴ Normal developmental milestones between three and nine years of age (Deonna, 1991);
- ⤴ Normal early language and psychomotor development;
- ⤴ Normal hearing tests and non-verbal skills; and
- ⤴ Usually unaffected intelligence levels.

Verbal auditory agnosia, or 'word deafness', is the most common feature and, usually, the first diagnosis is hearing loss (Rapin et al, 1977). This extends to familiar sounds like bells, whistles or a ringing telephone. Some children maintain their ability to write and to communicate through non-verbal methods (Gordon 1990, Soprano et al 1994).

With regard to acquired aphasia, its onset is insidious and progressive, with remissions and relapses. Stuttering may be a feature (Tutuncuoglu et al, 2002), along with mutism. Epileptic seizures are present in up to 80 per cent of cases, usually at night, which usually stop by the time the child becomes a teenager (Pauier et al, 1992).

The most common seizures are eyelid myoclonus, eye blinking, atypical absences, head drops and atonic fits of upper limbs, and automatism. Seizures may present before aphasia (Deonna et al, 1997).

### **Behavioural disorders**

Behavioural disorders are also associated with: irritability, hyperactivity, excitability, aggressiveness and depression. Psychotic or autistic features in severe cases may handicap children in understanding language (Deonna 1991, Klein et al 2000).

Some differential diagnoses include: autism, Rett's syndrome, Heller's syndrome, Angelman syndrome, hearing impairment, learning disability, BECTS, epilepsy, auditory/verbal processing disorder, attention deficit disorder, mental retardation, childhood schizophrenia or emotional/behavioural problems (Tuchman et al 1997, Tassinari et al 2002).

Diagnosis

EEG typically shows bilateral symmetrical and asymmetrical multifocal spikes and spike-waves over temporal and parieto-occipital lobes, which are correlated with persistence of language

impairment (Morell et al 1995, Fejerman et al 2000, Veggiotti et al 2002). It is postulated that EEG changes and seizures are related to abnormalities in speech areas (Holmes et al, 1981).

Magnetoencephalography often suggests that the intrasylvian cortex is a trigger of epileptic seizures in LKS (Sobet et al, 2000) and abnormal auditory evoked potentials are reported in these cases (Wioland et al, 2001), while single photon emission computed tomography (SPECT) and positron emission tomography (PET) reveal unilateral or bilateral changes in the temporal lobe (Da Silva et al, 1997).

Magnetic resonance imaging (MRI) and computed tomography (CT) brain scans are usually normal (Gascon et al 1990, Marescaux et al 1990). A neuro-psychology assessment is helpful to locate particular difficulties in a child and is important in planning educational needs, for monitoring progress and response to treatment.

Neurological assessment is usually normal, but neuropsychological evaluation is crucial to determine the nature of language disorder and intelligence level (Deonna, 1991).

The management of Landau-Kleffner syndrome involves:

1. Comprehensive history and neurological examination;
2. Comprehensive history of speech and language development;
3. Neuropsychological and speech assessment;
4. Psychoeducation of the patient and the family;
5. Behaviour therapy and family therapy to address anxiety and frustration; and
6. Occupational help and career guidance.

Speech therapy may be needed to increase the patient's communication skills. Children should also be seen and treated by a speech and language therapist and an educational psychologist to help with their speech and educational problems (Tharpe et al, 1991); With regard to antiepileptics, commonly used drugs include sodium valproate, ethosuximide, clonazepam and clobazam. Vigabatrin and felbamate are transiently effective (Appleton et al 1993, Fejerman et al 2000, Prats-Vinas 2002).

Sometimes a combination of two or three antiepileptics may also be used and if these drugs fail, steroid medications may be used for short periods.

Corticosteroids and intravenous gamma-globulins are efficacious treatments for clinical and EEG abnormalities (Tsuru et al 2000, Mikati et al 2002).

A highly specialised form of brain surgery called subpial transection has been tried, with some success (Solomon et al 1993, Nass et al 1993, Grote et al 1999).

## **Prognosis**

An early age of onset in patients (i.e. before they reach the age of four years), duration of aphasia for longer than one year, and duration and continuity of epilepsy with electrical status epilepticus

during sleep are linked with worse outcome (Bishop 1985, Rossi et al 1999) and aphasia is also likely to persist in adulthood (Matovani et al, 1980).

Although the outlook for seizures in children with Landau-Kleffner syndrome is good, many children will be left with significant language, learning and behavioural difficulties. The seizures usually stop by the age of 15 years, although one in 10 continues to have infrequent seizures (Mantovani et al, 1980).

Only 10 per cent to 20 per cent of children with LKS will have normal language and learning abilities in adulthood (Paquier et al, 1992). Complete recovery to permanent severe aphasia with remissions and relapses are noted (Mouridsen et al, 1995).

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