Autosomal dominant lateral temporal epilepsy

LGI1, cause autosomal dominant lateral temporal epilepsy, a genetic focal epilepsy syndrome.

Prevalence

Unknown but may be rare.

Age at onset

Typically in teenage years or early adult life; may start earlier, 5 to 10 years or later.

Sex

Males = females.

Neurological and mental state

Normal.

Genetics

Autosomal dominant inheritance with high (~80%) penetrance. It is the first non-ion channel idiopathic focal epilepsy to be described. Mutations of the leucine-rich, glioma-inacivated 1 (LGI1/Epitempin gene) on chromosome 10q are responsible for this disorder.

Clinical manifestations

Simple focal seizures that are the most common consist mainly of simple auditory hallucinations such as ringing, humming, clicking, or unspecified noises. These may infrequently progress to complex focal seizures and generalized tonic-clonic seizures (GTCS). Other sensory symptoms such as visual (lights, colors, and simple figures), olfactory, vertiginous, or cephalic are frequent with simple focal seizures while autonomic, mental, and motor symptoms are less common. Families with brief aphasic seizures have been described.

Secondarily GTCS are rare.

Timing

GTCS predominantly nocturnal.

Seizure-precipitating factors

Sleep.

Diagnostic procedures

MRI is normal.

Interictal EEG

Often normal; epileptiform abnormalities rarely occur.

Prognosis

Excellent. Patients are neurologically and mentally normal, and the condition does not appear to affect their otherwise normal life, particularly when on medication. Seizures are generally mild, infrequent, and well controlled with anti-epileptic medication.

Differential diagnosis

Symptomatic lateral temporal lobe epilepsy that lacks a similar family history and that is markedly different from hippocampal epilepsy and familial mesial temporal lobe epilepsy.

Management options

AEDs indicated for focal seizures. Response to carbamazepine is excellent.