Leucine-rich glioma-inactivated 1 (LGI1) antibody encephalitis is a recently described syndrome primarily characterized by seizures and cognitive decline. LGI1 antibody encephalitis may easily be mistaken for other neuropsychiatric syndromes, and clinical identification of the syndrome and confirmation with antibody testing is paramount, as EEG, MRI, and routine lumbar puncture studies may be unremarkable, particularly early in the clinical course. At the same time, early treatment with immunotherapy has been shown to favorably alter the prognosis. The annual incidence of LGI1 antibody encephalitis in the Netherlands has been estimated to be 0.83/million with a reported age range of 28 to 92 years and approximately equal sex distribution.\(^1,2\)

**Antibodies and Receptor**
Antibodies in this syndrome are related to the VGKC, specifically the LGI1 region of the VGKC channel complex.\(^3\) LGI1 is mainly expressed in the hippocampus and temporal cortex.\(^1\) LGI1 is part of an inhibitory pathway linking VGKC to AMPA receptors. Antibodies are detected by incubation of cerebrospinal fluid (CSF) or serum with human embryonic kidney cells, then using an antibody against human immunoglobulin G with fluorochrome or another chemical permitting microscopic detection.\(^1,3\) Testing of CSF and serum pairs is recommended, as LGI1 antibody has variably been reported as positive in one but negative in the other.\(^1,3,4\)

**Diagnosis**
Faciobrachial dystonic seizures (FBS) are the characteristic seizure type associated with this syndrome and are seen in 40% to 71% of LGI1 cases.\(^1,5\) FBS are not exclusively seen in LGI1 encephalitis but are relatively specific for it.\(^1\) FBS are involuntary contractions consisting of dystonic posture of the arm followed by facial contraction lasting 1 to 30 seconds, most commonly affecting a unilateral arm and the face.\(^1,2\) FBS are often under-recognized, but they may occur between eight and 200 times a day.\(^1,2\) The ipsilateral leg may be involved in up to 40% of cases, and these seizures may also be bilateral.\(^2\) FBS and subtle seizures generally precede the insidious development of cognitive decline and memory disturbance.\(^1,2\) Other seizure types can also be seen, including focal seizures with dyscognitive, autonomic motor, and gelastic features.\(^1\) In addition to memory disturbance, behavioral disturbances have also been described, including apathy, disinhibition, missing social cues, compulsive behavior, ego centricity, and spatial disorientation.\(^1\) Other associated symptoms that have been reported include insomnia, hyperhidrosis, and hyponatremia.\(^1\)

Imaging and laboratory testing may be unrevealing. MRI commonly but not always shows hippocampal T2 hyperintensity.\(^1\) MRI findings may be unilateral but are more commonly bilateral.\(^4\) MRI findings may be absent early on in the disease course.\(^2,4\) Most patients with prolonged symptoms showed medial temporal MRI changes.\(^4\) T1 hyperintensities in the basal ganglia have also been reported.\(^5\) Furthermore, FBS often have no EEG correlate, although longer seizures have been reported to have associated EEG findings.\(^1,2,5\) Finally, CSF pleocytosis may be seen, but in a reported case series was only seen in 23% of patients with encephalitis.\(^4\) Ultimately, up to 13% of patients who present with encephalopathy may be without MRI or CSF findings, consistent with encephalitis based upon reported data.\(^4\)

**Management**
The treatment of LGI1 antibody encephalitis is largely patterned after treatment to NMDA receptor encephalitis.\(^3\) First-line treatment consists of steroids followed by or in conjunction with plasma exchange/intravenous immunoglobulin.\(^3\) First-line treatment has been found to be effective in 80%.\(^1\) The median time from onset to nadir of illness was 22 weeks.\(^1\) Improvement is usually noted within 2 weeks of treatment, but time to recov-
LGI1 encephalitis has been misdiagnosed as Creutzfeldt-Jakob disease due to subacute cognitive decline associated with myoclonus and absent CSF pleocytosis. Other misdiagnoses include psychogenic movement disorder, paroxysmal kinesigenic dyskinesia, Stokes-Adams syndrome, and Alzheimer’s disease. Early diagnosis and treatment prior to the onset of cognitive decline will hopefully lead to better outcomes, as time to recovery has been correlated with time to initiation of immunotherapy, though not with time to initiation of AED. Furthermore, patients who presented with FBS and received no treatment or AED alone developed cognitive impairment, whereas patients who received immunotherapy did not develop cognitive impairment.

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