Seronegative Autoimmune Encephalitis

Only up to 44% of patients with AE have an antibody positive status. ‘Seronegative but suspected autoimmune encephalitis’ has received a consensus definition in 2013. The definition includes rapid progression of symptoms, along with exclusion of well-defined AE syndromes such as typical limbic encephalitis, absence of serum and CSF antibodies along with two of: MRI abnormalities suggestive of autoimmune encephalitis, CSF pleocytosis, CSF-specific oligoclonal bands or elevated CSF IgG index or brain biopsy showing inflammatory infiltrates, along with exclusion of other causes.

When to Suspect Autoimmune Encephalitis in Children?

The diagnosis of AE should be suspected in all children who develop a polysymptomatic syndrome encompassing encephalopathy, seizures, movement disorders, psychiatric features, gait disturbances and autonomic disturbances. The clinical features suggestive of autoimmune encephalitis include:

• Abrupt onset / rapid decline
• Autonomic instability
• Delirium slipping into catatonia and vice versa
• Urinary/ fecal incontinence
• Cognitive slowing • Gait and balance disorder
• Relapse after treatment for viral encephalitis
• Seizures that may be in the form of status epilepticus or multifocal drug resistant epilepsy or seizure clusters
• Involvement of multiple domains eg. Cognition and extrapyramidal system etc.
• CSF may also reveal features of inflammation in the absence of infection. Features that point away from the diagnosis of AIE include:

• A very chronic or indolent course
• Plateauing of symptoms
• No impairment in activities of daily living
• Cognition remaining intact
• Purely psychiatric symptoms

From: Paediatric autoimmune encephalopathies: clinical features, laboratory investigations and outcomes in patients with or without antibodies to known central nervous system autoantigens  https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3686256/