Pediatric Neurology: Chapter 129. Anti-NMDA receptor encephalitis in children: the disorder, its diagnosis, and treatment (Handbook of Clinical Neurology)

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Anti-NMDA receptor encephalitis is a newly characterized severe neuroautoimmune syndrome with a progressive, clinical course. Most often seen in females, it usually begins with a prodromal phase suggestive of an acute or subclinical upper respiratory tract infection that lasts for up to 2 weeks. This is followed by a psychotic and seizure phase in which the child may rapidly develop seizures, behavioral changes, and, less commonly in children, psychiatric symptoms, resulting in frequent misdiagnoses. The child may become mute and unresponsive but awake during the akinetic phase. Autonomic instability characterizes the hyperkinetic phase. A teratoma or, more rarely, another tumor type is found in 25% of affected adolescents beyond the first decade of life. The finding of oligoclonal protein electrophoresis (>80%) and antibodies in serum and cerebrospinal fluid directed against the NR1 subunit of the NMDA receptor confirms the diagnosis. Prognosis is improved with the appropriate use of immunosuppressant therapies. Relapses in children may be multiple and occur in 20–25% of cases. Recovery is slow and may take 3 years or longer. Even so, the child may not always regain its premorbid level of health.

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