



Pediatric Neurology: Chapter 130. Immune-mediated extrapyramidal movement disorders, including Sydenham chorea (Handbook of Clinical Neurology)

Russell C. Dale

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Immune-mediated extrapyramidal movement disorders typically occur in previously healthy children. Immune-mediated movement disorders may occur as a postinfectious, paraneoplastic, or idiopathic process. Sydenham chorea (SC) is the classical poststreptococcal movement and psychiatric disorder, and may be associated with other features of rheumatic fever. The outcome is typically good, although residual chorea, psychiatric disturbance, and relapses are possible. Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) is a syndrome of streptococcal-induced tics and obsessive-compulsive disorder. Although a number of investigators have reported an association between streptococcal infection and neuropsychiatric syndromes, the PANDAS hypothesis is controversial. Encephalitis lethargica is an encephalitic illness with parkinsonism, dyskinesias, and psychiatric disturbance as dominant features. The exact disease mechanism is not understood, although an autoimmune process is suspected. NMDA-R encephalitis is a new entity characterized by encephalitis with dramatic psychiatric disturbance, dyskinesias, cognitive alteration, and seizures. Patients have autoantibodies against the NMDA-R that appear to be pathogenic: immune therapies appear warranted to minimize disability. Movement disorders are also described associated with systemic lupus erythematosus and antiphospholipid syndrome. The differential diagnosis and investigation approach of acute-onset movement disorders are also discussed.

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