



Pediatric Neurology: Chapter 131. Systemic inflammatory and autoimmune disorders (Handbook of Clinical Neurology)

Daniela. Pohl, Susanne. Benseler

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Systemic disorders with possible involvement of the nervous system include a variety of diseases with presumed inflammatory and autoimmune pathomechanisms, among them Behçet disease, sarcoidosis, systemic lupus erythematosus, juvenile idiopathic arthritis, scleroderma, and Sjögren syndrome. This disease group encompasses systemic inflammatory disorders with a genetically defined dysregulation of the innate immune system as well as systemic autoimmune disorders characterized by alterations of the adaptive immunity such as autoantibodies and autoreactive T cells. Although more commonly diagnosed in adults, all of these diseases can manifest in childhood and some as early as infancy. Neurological involvement may represent the initial manifestation, and nearly every neurological symptom can be caused by inflammatory/autoimmune diseases. In a child with (sub)acute onset of otherwise unexplained neurological findings, consideration of inflammatory/autoimmune disorders may be of crucial therapeutic and prognostic importance. In the absence of disease-specific clinical features, the initial diagnostic workup is broad. Basic blood tests include inflammatory markers and autoantibodies. Cerebral magnetic resonance imaging and a lumbar puncture with measurement of opening pressure as well as cerebrospinal fluid analysis are indicated in most patients with central nervous system (CNS) involvement. Skin, muscle, or organ biopsies (e.g., renal) may provide additional information. Especially in patients with isolated CNS involvement, a brain biopsy may be indicated. Timely recognition and treatment of CNS inflammation may improve or even reverse clinical symptoms and prevent secondary brain injury.

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