

Pediatric Neurology: Chapter 134. Effect of autoimmune diseases on cognitive function (Handbook of Clinical Neurology)

Lily Lim, Sarah Lippe, Earl Silverman



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The location of both autoimmune processes and other causes of brain inflammation is important in determining the impact of inflammation on brain function. This chapter focuses on autoimmune and infectious diseases leading to inflammatory brain disease resulting in cognitive defects with a special focus on systemic lupus erythematosus (SLE). Collectively called neuropsychiatric SLE (NPSLE), NPSLE occurs in 20–95% of pediatric patients with SLE (pSLE). The incidence of cognitive dysfunction is difficult to ascertain in pediatric patients as few studies have been performed. Using formal neurocognitive testing of unselected pediatric SLE patients, the rate of cognitive abnormalities was approximately 50% and impairment was associated with longer disease duration in one study. A second small study showed global depression on performance and academic scores while a larger study using a neuropsychiatric inventory showed a 55% rate of dysfunction. These diverging findings may result from the lack of a standardized cognitive assessment battery. The Childhood Arthritis and Rheumatology Research Alliance (CARRA) group of pediatric rheumatologists proposed a 2 hour 40 minutes battery for assessment of cognitive testing of SLE patients from age 9 to 18 years. Further assessments using this battery should provide a better neurocognitive profile of pSLE.

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