



Pediatric Neurology Part I: Chapter 63. Infantile spasms (Handbook of Clinical Neurology)

Richard A. Hrachovy, James D. Frost

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Infantile spasms are a unique disorder of infancy and early childhood. The average age at onset of infantile spasms is 6 months and the average incidence of the disorder is approximately 0.31 per 1000 live births. Approximately one-quarter of patients will spontaneously stop having spasms within 1 year of onset. There are three main types of epileptic spasms: flexor, extensor, and mixed flexor-extensor. Spasms frequently occur in clusters and commonly occur upon arousal from sleep. The motor spasms are frequently confused with other normal and abnormal infant behaviors. Typically, the interictal EEG reveals hypsarrhythmia or one of its variants. A variety of ictal EEG patterns may be seen, the most common of which is a generalized slow-wave transient followed by an attenuation of the background activity in all regions. The primary treatment objective is to improve the EEG and stop the spasms as soon as possible and to avoid prolonged treatment durations with any form of therapy. Currently, there is no conclusive evidence that medical or surgical treatment of infantile spasms significantly alters long-term outcome. Although the pathophysiological mechanism underlying infantile spasms is unknown, several animal models of infantile spasms have been developed in recent years

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