Pediatric Neurology: Chapter 85. Alternating hemiplegia of childhood (Handbook of Clinical Neurology)

Sujay Kansagra, Mohamad A. Mikati, Federico Vigevano

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Alternating hemiplegia of childhood (AHC) is a very rare disease characterized by recurrent attacks of loss of muscular tone resulting in hypomobility of one side of the body. The etiology of the disease due to ATP1A3 gene mutations in the majority of patients. Few familial cases have been described. AHC has an onset in the first few months of life. Hemiplegic episodes are often accompanied by other paroxysmal manifestations, such as lateral eyes and head deviation toward the hemiplegic side and a very peculiar monocular nystagmus. As the attack progresses, hemiplegia can shift to the other side of the body. Sometimes the attack can provoke bilateral paralysis, and these patients may have severe clinical impairment, with difficulty in swallowing and breathing. Hemiplegic attacks may be triggered by different stimuli, like bath in warm water, motor activity, or emotion. The frequency of attacks is high, usually several in a month or in a week. The duration is variable from a few minutes to several hours or even days. Sleep can stop the attack. Movement disorders such as dystonia and abnormal movements are frequent. Cognitive delay of variable degree is a common feature. Epilepsy has been reported in 50% of the cases, but seizure onset is usually during the third or fourth year of life. Many drugs have been used in AHC with very few results. Flunarizine has the most supportive anecdotal evidence regarding efficacy.

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