عنوان مقاله:

Hyperimmunoglobulin-D Syndrome in Children: A Review Article

محل انتشار:

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خلاصه مقاله:

Hyperimmunoglobulin-D syndrome (HIDS) is a rare, autosomal recessively inherited autoinflammatory disease caused by mutations in the mevalonate kinase gene. HIDS usually starts in infancy with recurrent fever episodes lasting three to seven days and recurring every three to six weeks, with only partial symptom decrease in adulthood. Fever is typically accompanied by abdominal pain, vomiting, diarrhea and cervical lymphadenopathy, and sometimes by skin and joint symptoms. Blood leukocytes and serum C-reactive protein (CRP) are elevated during the episode, and in addition, high levels of interleukine-۱ (IL-۱), IL-۶ and tumor necrosis factor (TNF) and respective soluble receptors are measured. Currently, there is no established treatment for HIDS. So far, four children have been successfully treated by TNF-alpha inhibitor (etanercept) and three children with IL-۱ receptor antagonist (anakinra). The current study is a narrative review about the updates of HIDS.

کلمات کلیدی:

Hyper IgD Syndrome, Mevalonate Kinase Deficiency, Child

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