

Periodic Fever and Maculopapular Rash

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Chapter

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Abstract

Periodic fever syndrome with hyper-IgD syndrome (HIDS), is an autosomal recessive autoinflammatory disease characterized by recurrent episodes of fever, abdominal pain, diarrhea, rash, arthralgia, aphthous ulcers, and lymphadenopathy with systemic inflammation

Serum IgD is increased in most patients with HIDS, while more than 20% of them have normal values of serum IgD and almost 50% of patients with other autoinflammatory disorders are identified to have increased levels of serum IgD

The most appropriate laboratory analysis for diagnostic of HIDS is testing of mevalonic acid level in urine during fever episodes

Keywords

Hyper-IgD syndrome MVK Urine mevalonic acid

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