

[Pediatr Allergy Immunol.](#) 2002 Apr;13(2):140-2.

Prolidase deficiency with hyperimmunoglobulin E: a case report.

[Lopes I](#), [Marques L](#), [Neves E](#), [Silva A](#), [Taveira M](#), [Pena R](#), [Vilarinho L](#), [Martins E](#).

Source

Instituto de Genética Médica, Hospital Crianças Maria Pia, Porto, Portugal.

Abstract

Prolidase deficiency is a rare, inherited disorder characterized by ulceration of the skin, mental retardation, and massive urinary excretion of imidodipeptides. Most patients also have recurrent infections, an unusual facial appearance, and splenomegaly. We describe a girl presenting with chronic dermatitis, recurrent respiratory tract infections since her first months of life, and facial features characteristic of prolidase deficiency. The diagnosis of prolidase deficiency was made at 4.5 months of age. The immunologic study in this patient showed an extreme and progressive increase of total immunoglobulin E (IgE) in serum (reaching the value of 77,600 IU/l) and defective chemotactic function of the neutrophils. Treatment with a hyper-proteic diet supplemented with ascorbic acid, manganese chlorite, and topical proline resulted in reduction of the frequency and severity of the infections and significant improvement of the skin lesions. The authors discuss the immunologic alterations and the favorable evolution with treatment in this patient.

PMID:

12000488

[PubMed - indexed for MEDLINE]