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## DERMATITIS HERPETIFORMIS: Skin Manifestation of Celiac Disease

**D**ermatitis herpetiformis (DH) is a chronic, intensely itchy, blistering skin manifestation of gluten sensitive enteropathy, commonly known as celiac disease. DH affects 15 to 25 percent of people with celiac disease.<sup>1</sup> Most people with DH have no other symptoms of celiac disease. DH is found mainly in adults and is more common in men and people of northern European descent.



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### SYMPTOMS

DH is characterized by **small, clustered papules and vesicles** that erupt symmetrically on the elbows, knees, buttocks, back, or scalp. Men may also have oral or genital lesions. A burning sensation may precede lesion formation. Lesions are usually scratched off by the time a patient comes in for a physical exam.

### CAUSES

DH is caused by the deposit of immunoglobulin A (IgA) in the skin, which triggers further immunologic reactions resulting in lesion formation. DH is an external manifestation of an abnormal immune response to gluten, in which IgA antibodies form against the skin antigen epidermal transglutaminase.

### DIAGNOSIS

A skin **biopsy** is the first step in diagnosing DH. Direct immunofluorescence of clinically normal skin adjacent to a lesion shows granular IgA deposits in the upper dermis. Histology of lesional skin may show microabscesses containing neutrophils and eosinophils but may reveal only excoriation due to the intense itching patients experience.

Skin biopsies performed on the affected skin are nearly always positive for IgA deposition.<sup>2</sup> Blood tests for antiendomysial or anti-tissue transglutaminase antibodies may also suggest celiac disease.

A positive biopsy and serology confirm celiac disease. In the absence of these results, patients should be referred to a gastroenterologist for a definitive diagnosis via intestinal biopsy.<sup>3</sup>

<sup>2</sup>Zone JJ. Skin manifestations of celiac disease. *Gastroenterology*. 2005;128:s87–s91.

<sup>3</sup>Abenavoli L, Proietti I, Leggio L, et al. Cutaneous manifestations in celiac disease. *World Journal of Gastroenterology*. 2006;12(6):843–852.

<sup>1</sup>Rodrigo L. Celiac disease. *World Journal of Gastroenterology*. 2006;12(41):6585–6593.

## TREATMENT

**Dapsone**, a sulfone, provides immediate relief of symptoms. For patients who cannot tolerate dapsone, sulfapyridine or sulfamethoxyypyridazine may be used, although these drugs are less effective than dapsone. A strict gluten-free diet is the only treatment for the underlying disease. Even with a gluten-free diet, drug therapy may need to be continued for 1 to 2 years.<sup>4</sup>

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## THE CELIAC DISEASE AWARENESS CAMPAIGN

The National Institutes of Health Celiac Disease Awareness Campaign provides current, comprehensive, science-based information about the symptoms, diagnosis, and treatment of celiac disease, also known as celiac sprue, nontropical sprue, and gluten-sensitive enteropathy. The Awareness Campaign is an initiative of the National Digestive Diseases Information Clearinghouse, a service of the National Institute of Diabetes and Digestive and Kidney Diseases.

Download this publication and learn more about the Awareness Campaign at [www.celiac.nih.gov](http://www.celiac.nih.gov).

Celiac Disease Awareness Campaign  
c/o National Digestive Diseases Information Clearinghouse  
2 Information Way  
Bethesda, MD 20892-3570  
Phone: 1-800-891-5389  
TTY: 1-866-569-1162  
Fax: 703-738-4929  
Email: [nddic@info.niddk.nih.gov](mailto:nddic@info.niddk.nih.gov)  
Internet: [www.digestive.niddk.nih.gov](http://www.digestive.niddk.nih.gov)

The National Digestive Diseases Information Clearinghouse (NDDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). The NIDDK is part of the National Institutes of Health of the U.S. Department of Health and Human Services. Established in 1980, the Clearinghouse provides information about digestive diseases to people with digestive disorders and to their families, health care professionals, and the public. The NDDIC answers inquiries, develops and distributes publications, and works closely with professional and patient organizations and Government agencies to coordinate resources about digestive diseases.

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<sup>4</sup>Nicolas MEO, Krause PK, Gibson LE, Murray JA. Dermatitis herpetiformis. *International Journal of Dermatology*. 2003;42:588-600.