Celiac Disease
AWARENESS CAMPAIGN
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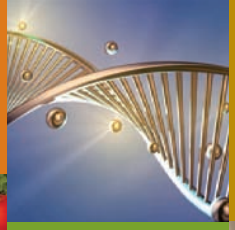
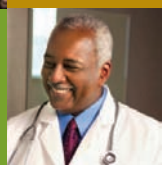
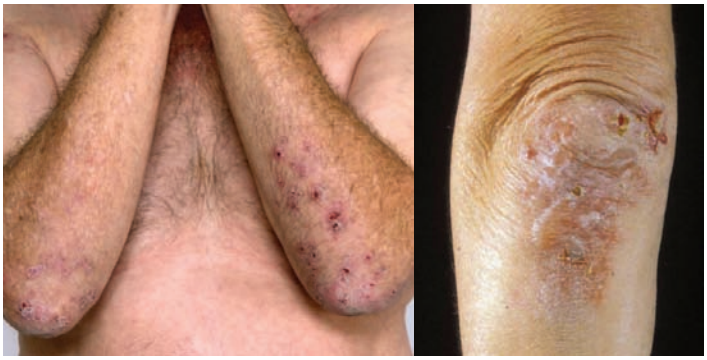




PROVIDER *Points*

DERMATITIS HERPETIFORMIS:
Skin Manifestation of Celiac Disease

Dermatitis herpetiformis (DH) is a chronic, intensely itchy, blistering skin manifestation of gluten-sensitive enteropathy, commonly known as celiac disease. DH is a rash that affects about 10 percent of people with celiac disease.[1] DH is found mainly in adults and is more common in men and people of northern European descent; DH is rarely found in African Americans and Asian Americans.[2]



SYMPTOMS

Dermatitis herpetiformis is characterized by small, clustered papules and vesicles that erupt symmetrically on the elbows, knees, buttocks, back, or scalp. The face and groin can also be involved. A burning sensation may precede lesion formation. Lesions are usually scratched off by the time a patient comes in for a physical exam, and the rash may appear as erosions and excoriations.

Patients with DH may also experience dental enamel defects to permanent teeth, which is another manifestation of celiac disease. Less than 20 percent of people with DH have symptoms of celiac disease.[3]

CAUSES

Dermatitis herpetiformis 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.

Family studies show that 5 percent of first-degree relatives of a person with DH will also have DH. An additional 5 percent of first-degree relatives of a person with DH will have celiac disease.[4] Various other autoimmune diseases are associated with DH, the most common being hypothyroidism.

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. However, histology may reveal only excoriation due to the intense itching that patients experience.

Blood tests for antiendomysial or anti-tissue transglutaminase antibodies may also suggest celiac disease. Blood tests for epidermal transglutaminase antibodies are positive in more than 90 percent of cases.[5] All of these tests will become negative with prolonged adherence to a gluten-free diet.

A positive biopsy and serology confirm DH and should be taken as indirect evidence of small bowel damage. A biopsy of the small bowel is usually not needed for DH diagnosis. However, if clinical signs of gastrointestinal disease are evident on examination, further workup may be required.[2] Whether or not intestinal damage is evident, a gluten-free diet should be implemented because the rash of DH is gluten sensitive.[4]

TREATMENT

The sulfone dapsone can provide immediate relief of symptoms. For patients who cannot tolerate dapsone, sulfapyridine or sulfamethoxypyridazine may be used, although these medications are less effective than dapsone. A strict gluten-free diet is the only treatment for the underlying disease. Even with a gluten-free diet, medication therapy may need to be continued from a few months to 2 years.

DH can go into remission, which is defined as absence of skin lesions and symptoms of DH for more than 2 years while not taking sulfones or other treatments and not adhering to a gluten-free diet. Cohort studies showing DH remission provide support for reducing sulfone therapy and weaning from a gluten-free diet in patients with well-controlled DH.[6]

REFERENCES

1. Ruiz AR. Celiac sprue. The Merck Manual website. www.merckmanuals.com/professional/gastrointestinal_disorders/malabsorption_syndromes/celiac_disease.html. Updated August 2012. Accessed December 4, 2013.
2. Bolotin D, Petronic-Rosic V. Dermatitis herpetiformis: part II. Diagnosis, management, and prognosis. *Journal of the American Academy of Dermatology*. 2011;64(6):1027–1033.
3. Alonzo-Llamazares J, Gibson LE, Rogers RS. Clinical, pathologic, and immunopathologic features of dermatitis herpetiformis: review of the Mayo Clinic experience. *International Journal of Dermatology*. 2007;46(9):910–919.
4. Sapone A, Bai JC, Ciacci C, et al. Spectrum of gluten-related disorders: consensus on new nomenclature and classification. BioMed Central website. www.biomedcentral.com/1741-7015/10/13. Published February 7, 2012. Accessed July 26, 2013.
5. Rose C, Armbruster FP, Ruppert J, Igl B-W, Zillikens D, Shimanovich I. Autoantibodies against epidermal transglutaminase are a sensitive diagnostic marker in patients with dermatitis herpetiformis on a normal or gluten-free diet. *Journal of the American Academy of Dermatology*. 2009;61(1):39–43.
6. Paek SY, Steinberg SM, Katz SI. Remission in dermatitis herpetiformis: a cohort study. *Archives of Dermatology*. 2011;147(3):301–305.

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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.*

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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.

This publication may contain information about medications and, when taken as prescribed, the conditions they treat. When prepared, this publication included the most current information available. For updates or for questions about any medications, contact the U.S. Food and Drug Administration toll-free at 1–888–INFO–FDA (1–800–463–6332) or visit *www.fda.gov.* Consult your health care provider for more information.

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