Celiac Disease

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INTRODUCTION

- A small bowel disorder characterized by mucosal inflammation, villous atrophy, and crypt hyperplasia, which occurs upon exposure to dietary gluten and which demonstrates improvement after withdrawal of gluten from the diet.
- Celiac disease (also called gluten-sensitive enteropathy and non tropical sprue) was first described by Samuel Gee in 1888

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CLASSIFICATION

- Classic disease
- Atypical celiac disease
- Asymptomatic (silent) celiac disease
- Latent celiac disease

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Classic disease

- The classic celiac disease or gluten-sensitive enteropathy includes the following three features:
  - villous atrophy;
  - symptoms of malabsorption such as steatorrhea, weight loss, or other signs of nutrient or vitamin deficiency
  - resolution of the mucosal lesions and symptoms upon withdrawal of gluten-containing foods, usually within a few weeks to months.

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Atypical celiac disease

Patients with atypical disease exhibit minor gastrointestinal complaints. They can display anemia, dental enamel defects, osteoporosis, arthritis, increased transaminases, neurological symptoms, or infertility.

However, most of these patients show severe mucosal damage and possess the celiac specific antibody pattern.

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Asymptomatic (silent) celiac disease

- Patients are often recognized incidentally based upon screenings for antibodies against gliadin or tissue transglutaminase.

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Latent celiac disease

- Celiac disease was present before, usually in childhood; the patient recovered completely with a gluten-free diet, remaining "silent" even when a normal diet was reintroduced
- A normal mucosa was diagnosed at an earlier occasion while ingesting a normal diet, but celiac disease developed later.

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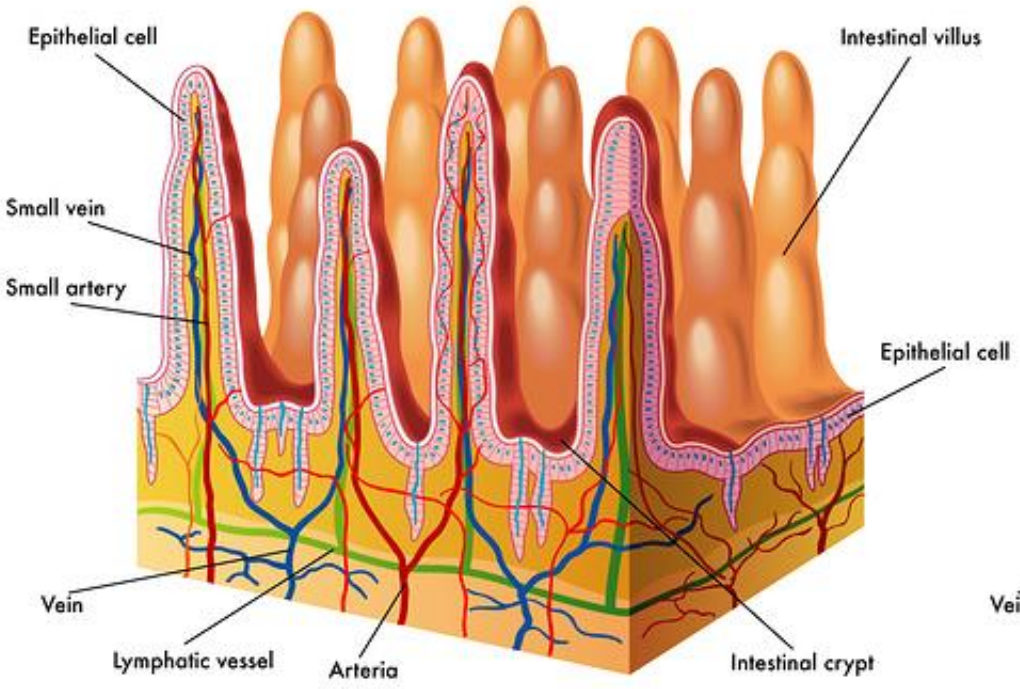
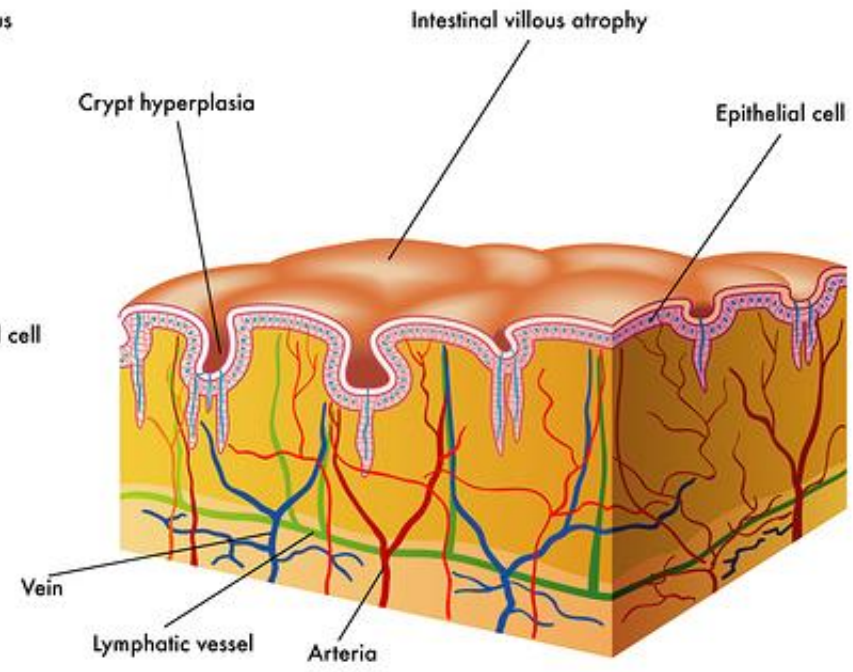
NORMAL



CELIAC DISEASE



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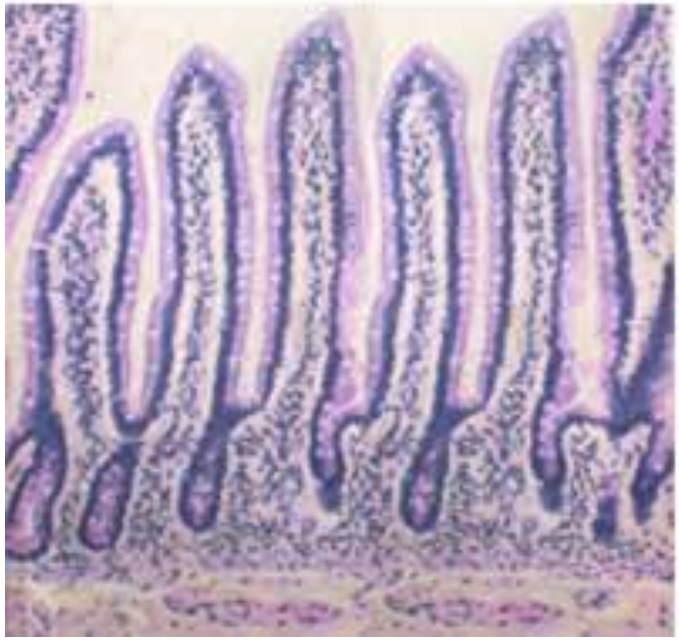
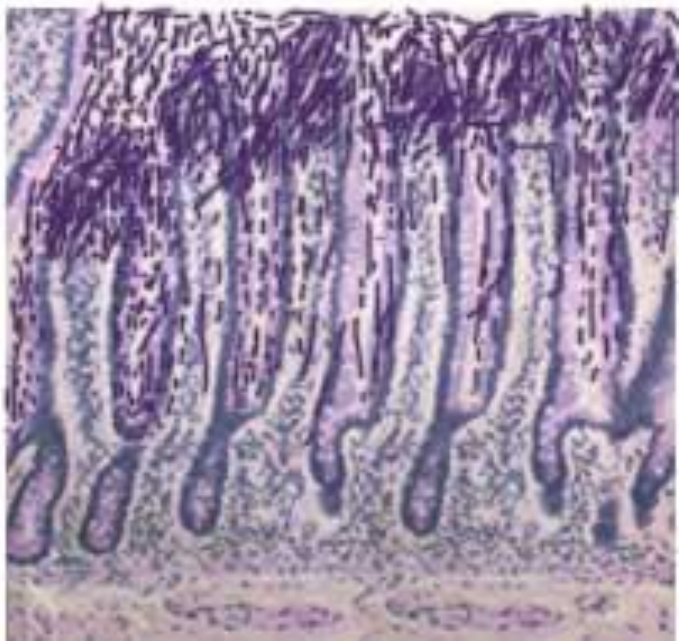
Healthy normal villi of the small intestine(as seen under the microscope)



Damaged villi of the small intestine



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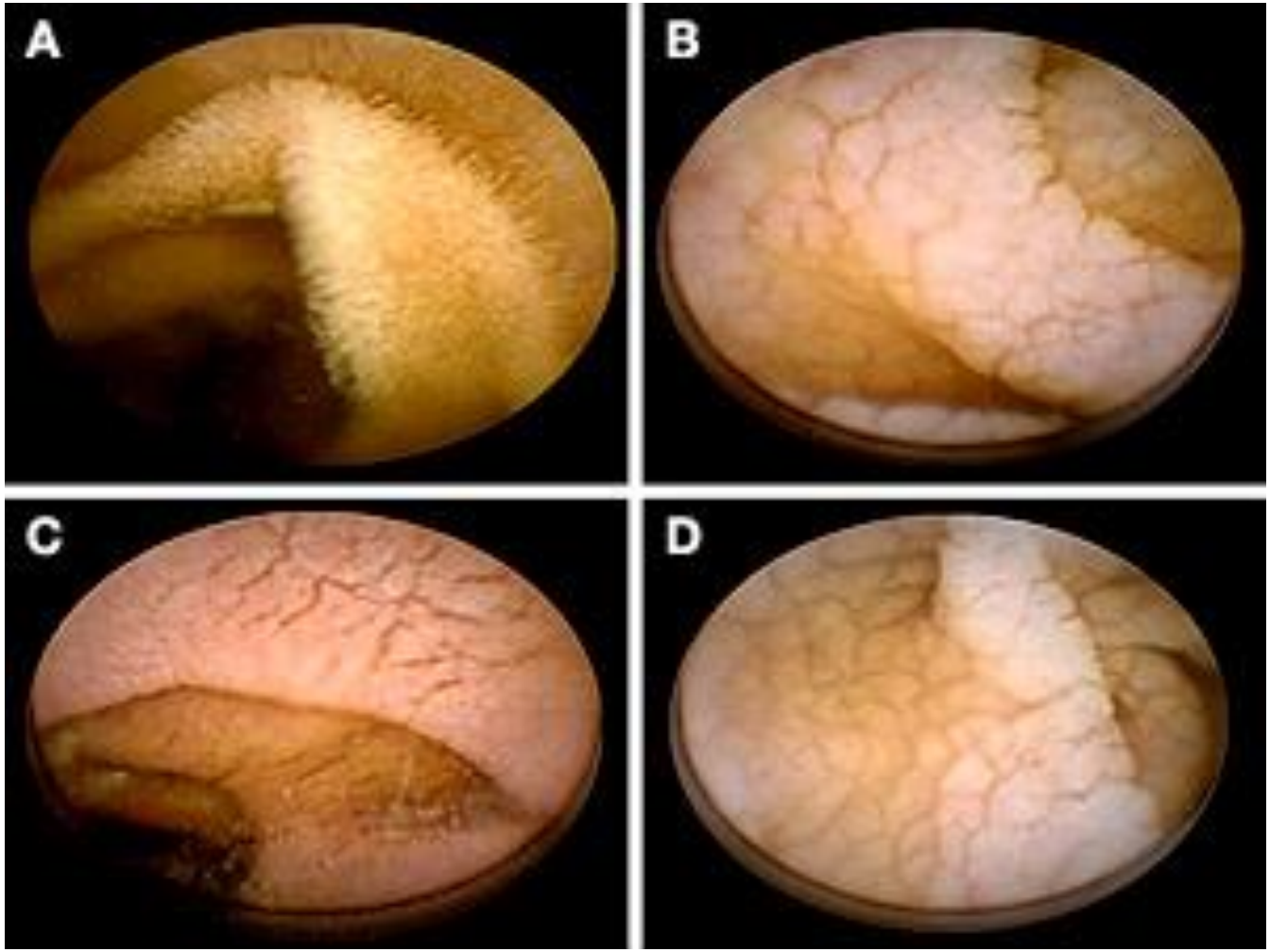

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EPIDEMIOLOGY

- Epidemiological studies in Northern European ancestry with blood tests with biopsy verification have reported prevalences of 1:70 to 1:300 in most countries

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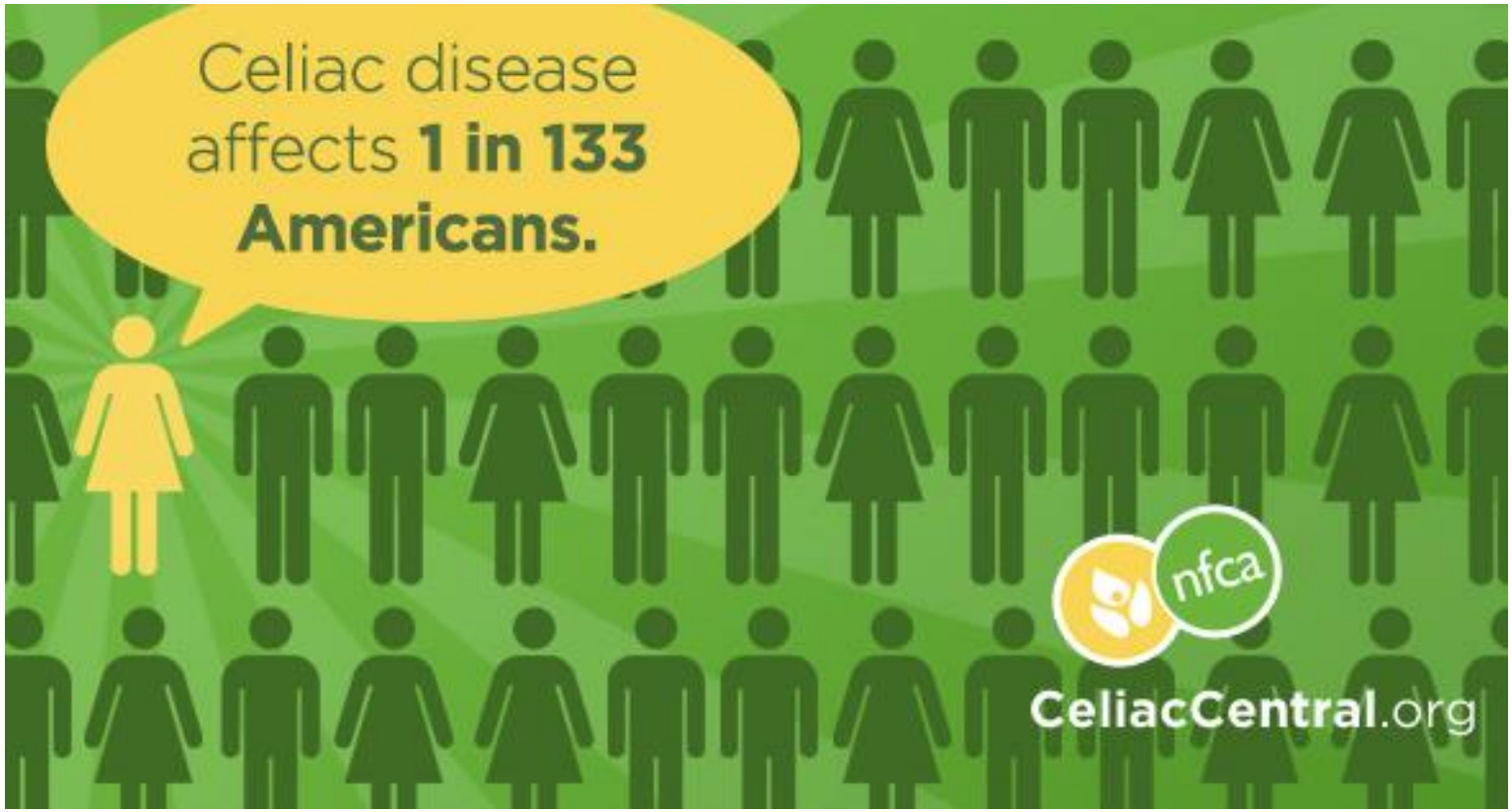

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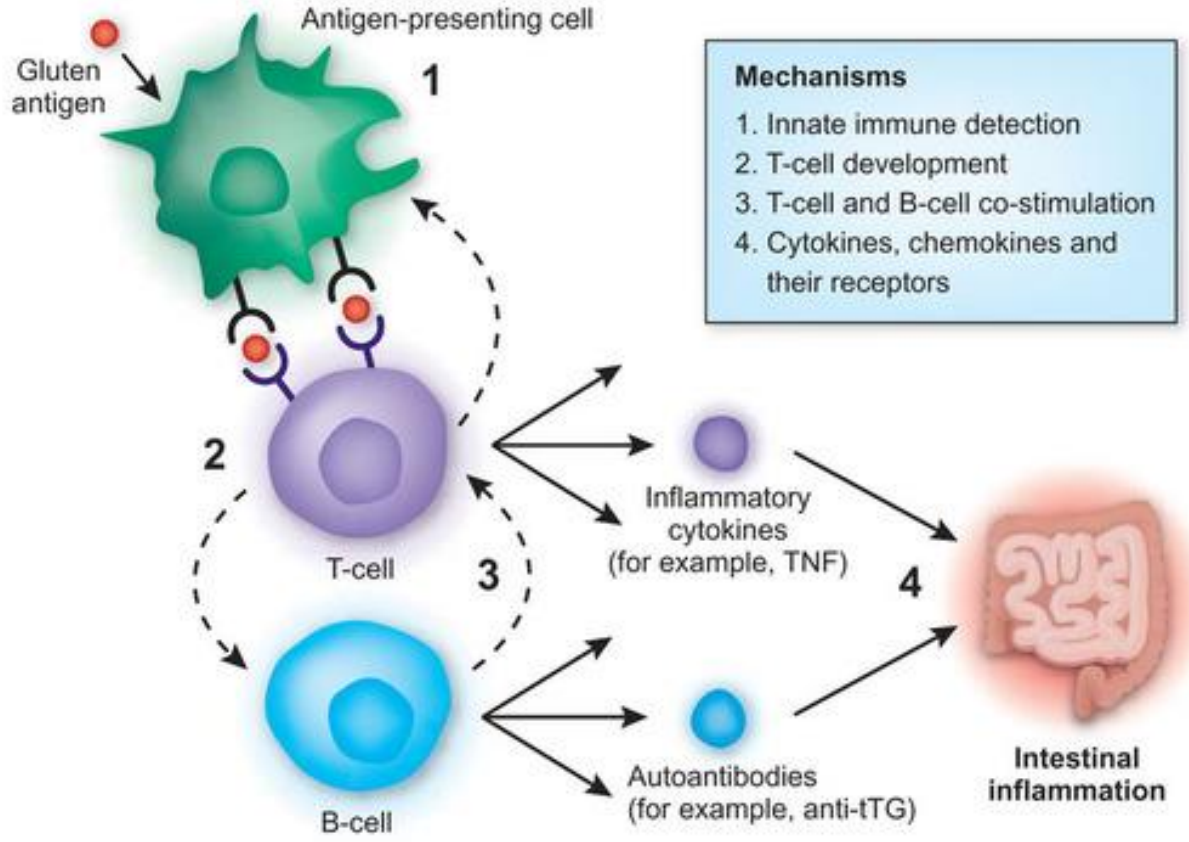
PATHOGENESIS

- Genetic factors
  - The frequent intrafamilial occurrence and the remarkably close association with the HLA-DQ2 and/or DQ8 gene loci provides the basis of our current understanding of celiac disease
  - HLA typing for DQ2 (DQA1*05; DQB1*02) and DQ8 (DQA1*03; DQB1*0302) may be useful in individuals with equivocal small bowel histologic findings since celiac disease is unlikely if neither is present
  - Homozygosity for HLA DQ2 has been associated with an increased risk for celiac disease and enteropathy-associated T-cell lymphoma
- Serum autoantibodies
- Gliadin reactive T cells



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CLINICAL MANIFESTATIONS

- Gastrointestinal manifestations
- Non gastrointestinal manifestations
  - Neuropsychiatric disease
  - Arthritis
  - Iron deficiency
  - Metabolic bone disease
  - Hyposplenism
  - Kidney disease
  - Idiopathic pulmonary hemosiderosis

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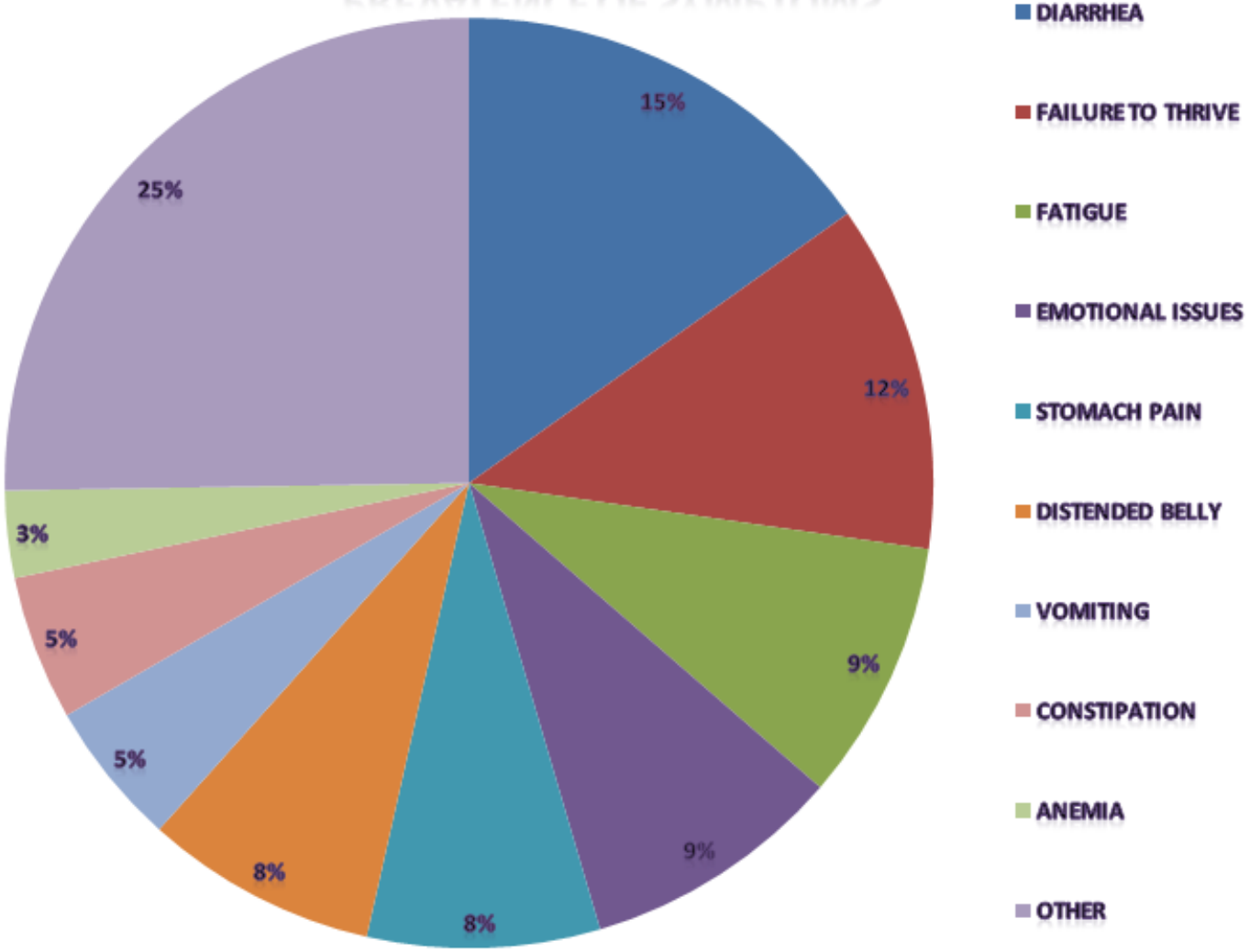
PREVALENCE OF SYMPTOMS



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WHO SHOULD BE TESTED?

- Those with gastrointestinal symptoms including
  - Chronic or recurrent diarrhea,
  - Malabsorption, weight loss,
  - Abdominal distension or bloating.
  - Irritable bowel syndrome or severe lactose intolerance.

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WHO SHOULD BE TESTED?

- Individuals without other explanations for signs and symptoms such as iron deficiency anemia, folate or vitamin B12 deficiency, persistent elevation in serum ALT, short stature, delayed puberty, recurrent fetal loss, low birthweight infants, reduced fertility, persistent aphthous stomatitis, dental enamel hypoplasia, idiopathic peripheral neuropathy, nonhereditary cerebellar ataxia, or recurrent migraine headaches.
- Patients with type 1 diabetes mellitus, and first-degree relatives of individuals with celiac disease if they have signs, symptoms, or laboratory evidence of possible celiac disease

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CELIAC DISEASE DIAGNOSIS

- Blood tests
- Small intestinal biopsy

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SERUM ANTIBODY ASSAYS

- IgA endomysial antibody (IgA EMA)
- IgA tissue transglutaminase antibody (IgA tTG)
- IgG tissue transglutaminase antibody (IgG tTG)
- IgA deamidated gliadin peptide (IgA DGP)
- IgG deamidated gliadin peptide (IgG DGP)

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Assay sensitivity and specificity

- IgA endomysial antibodies – sensitivity 85 to 98 % ; specificity 97 to 100 %
- IgA tissue transglutaminase antibodies – sensitivity 90 to 98 %; specificity 95 to 97 %
- IgA deamidated gliadin peptide – sensitivity 94 %; specificity 99 %
- IgG deamidated gliadin peptide – sensitivity 92 %; specificity 100 %

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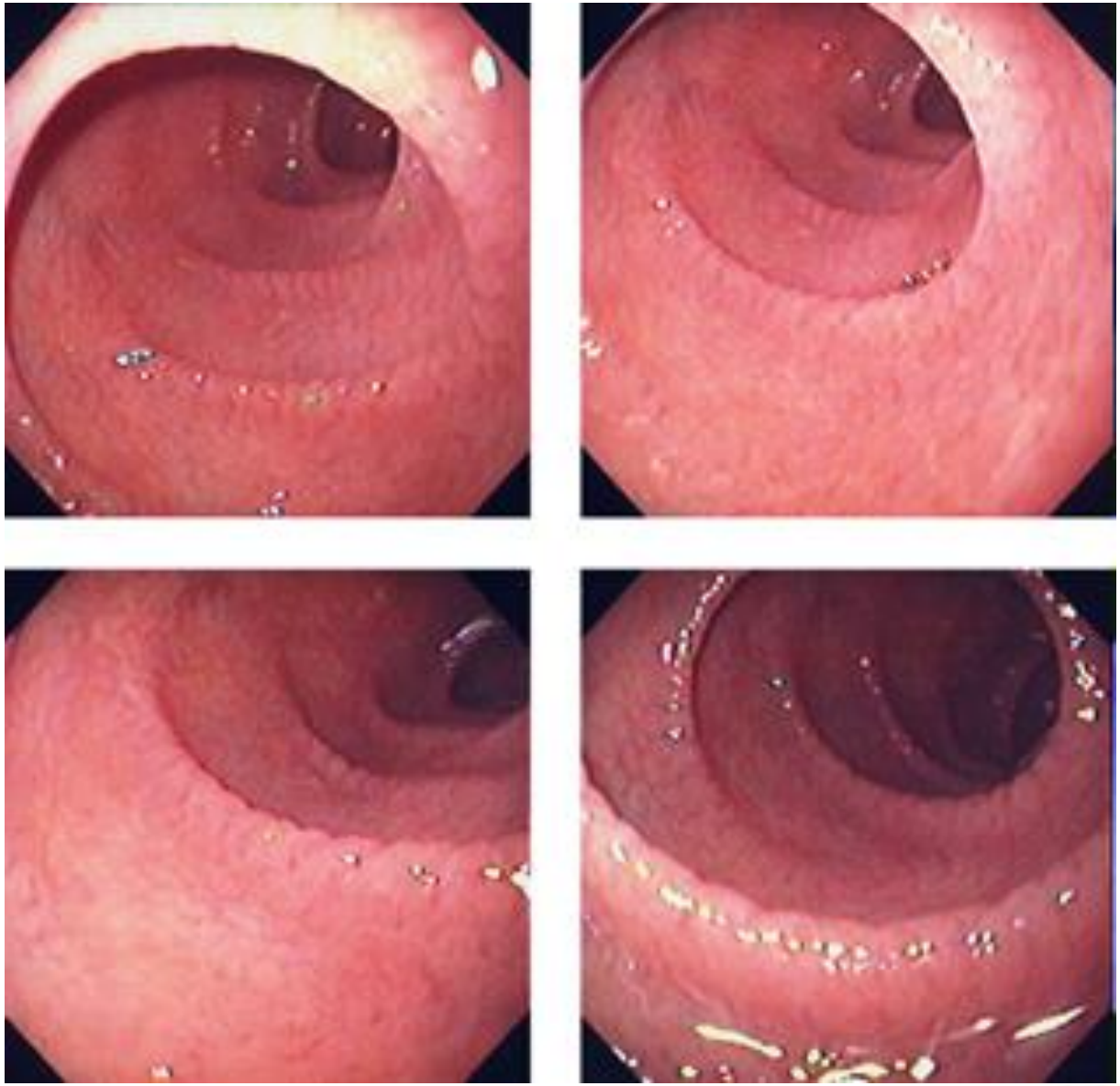

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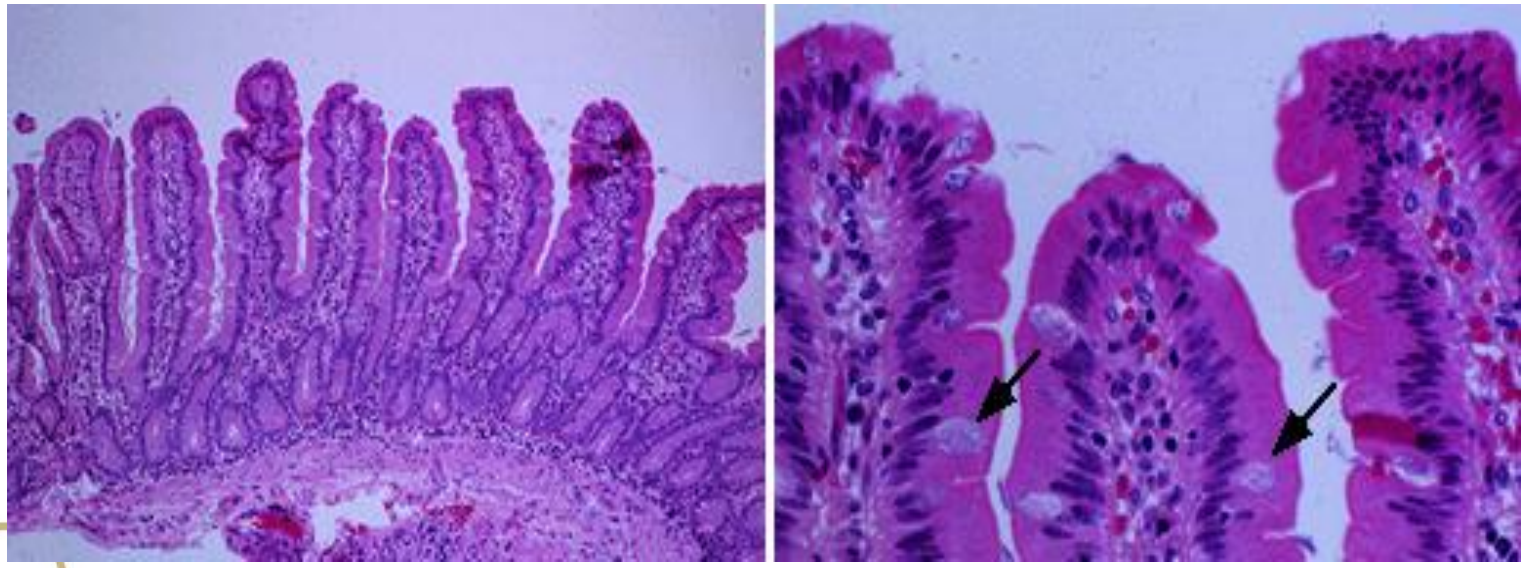
Normal Mucosa



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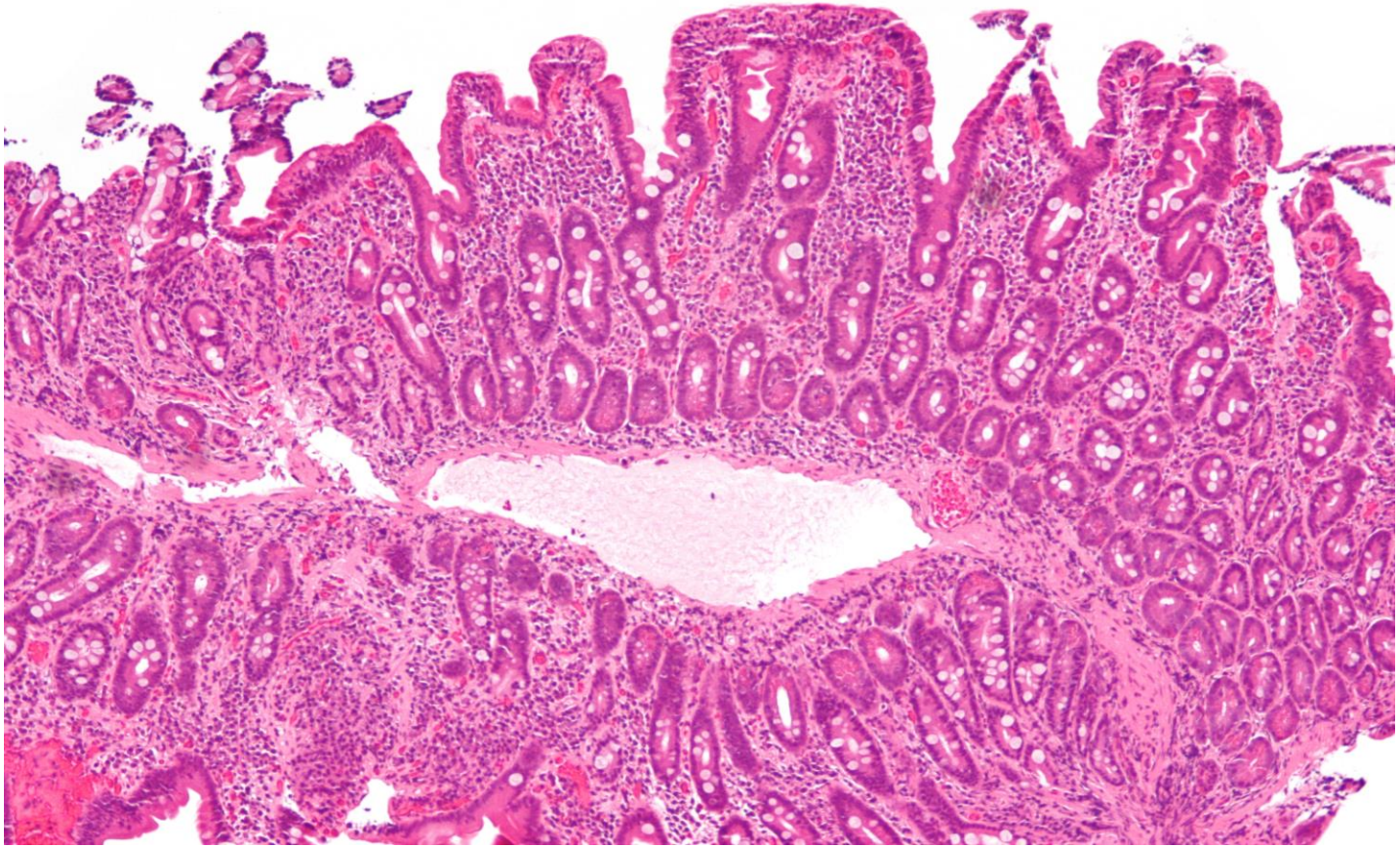
Celiac Disease



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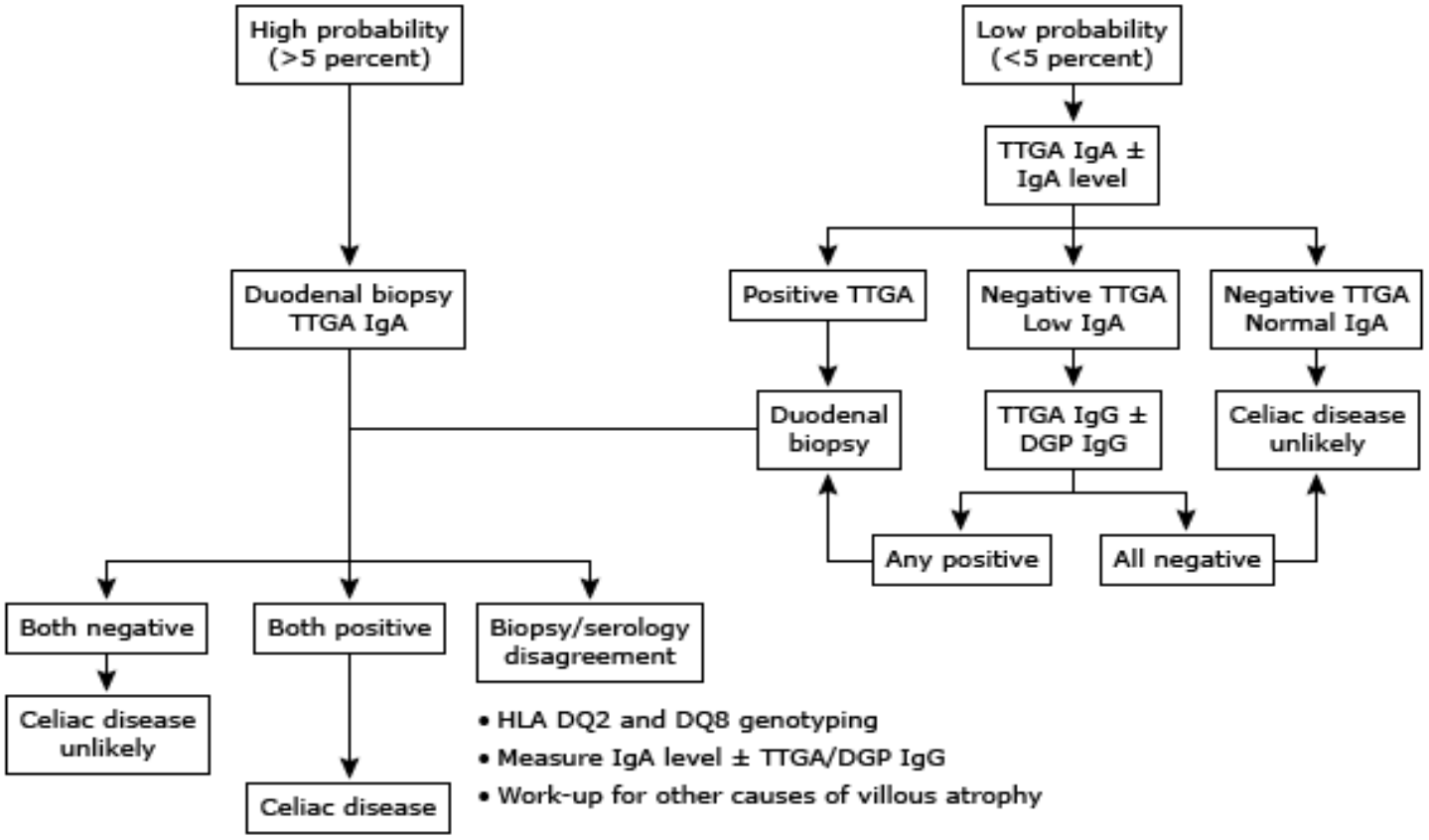
Celiac disease testing algorithm



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Management of celiac disease

- Consultation with a skilled dietitian
- Education about the disease
- Lifelong adherence to a gluten-free diet
- Identification and treatment of nutritional deficiencies
- Access to an advocacy group
- Continuous long-term follow-up by a multidisciplinary team

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CELIAC DISEASE COMPLICATIONS

- Nonresponsive celiac disease
- Refractory celiac disease
- Ulcerative jejunitis
- Lymphoma
- Skin conditions

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Nonresponsive celiac disease

- Approximately 5 percent of individuals
  - Patients with poor compliance or inadvertent gluten ingestion
  - Patients with clinical or histologic features that overlap with celiac disease but are caused by other disorders
  - Patients with concurrent disorders
  - Patients with refractory sprue
  - Patients with ulcerative jejunitis or intestinal lymphoma

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Foods and products that may contain gluten

Frequently overlooked foods that may contain gluten and need to be verified:
Brown rice syrup
Breading and coating mixes
Croutons
Energy bars
Flour or cereal products
Imitation bacon
Imitation seafood
Marinades
Panko (Japanese bread crumbs)
Pastas
Processed luncheon meats
Sauces, gravies
Self-basting poultry
Soy sauce or soy sauce solids
Soup bases
Stuffings, dressing
Thickeners (roux)
Communion wafers
Herbal supplements
Drugs and over-the-counter medications
Nutritional supplements
Vitamins and mineral supplements
Play-dough, crayons, paint, glue, paper mache: A potential problem if the child puts their hands on or in the mouth while playing. Wash hands after using these products.

NOT ALLOWED in any form:
Wheat (einkorn, durum, faro, graham, kamut, semolina, spelt)
Rye
Barley
Triticale
Malt, malt flavoring, malt vinegar (are generally made from barley, verify the source)

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Refractory sprue

- **Type 1:**
  - Normal population of intraepithelial lymphocytes.
- **Type 2:**
  - Aberrant or premalignant population of intraepithelial lymphocytes based upon clonality analysis of T-cell receptors and immunophenotyping.
  - Type 2 can progress to enteropathy-associated T-cell lymphoma, which may present clinically as ulcerative jejunitis. The diagnosis can be established on biopsy; CT, MRI, and 18F-FDG PET scans can help identify suspicious areas

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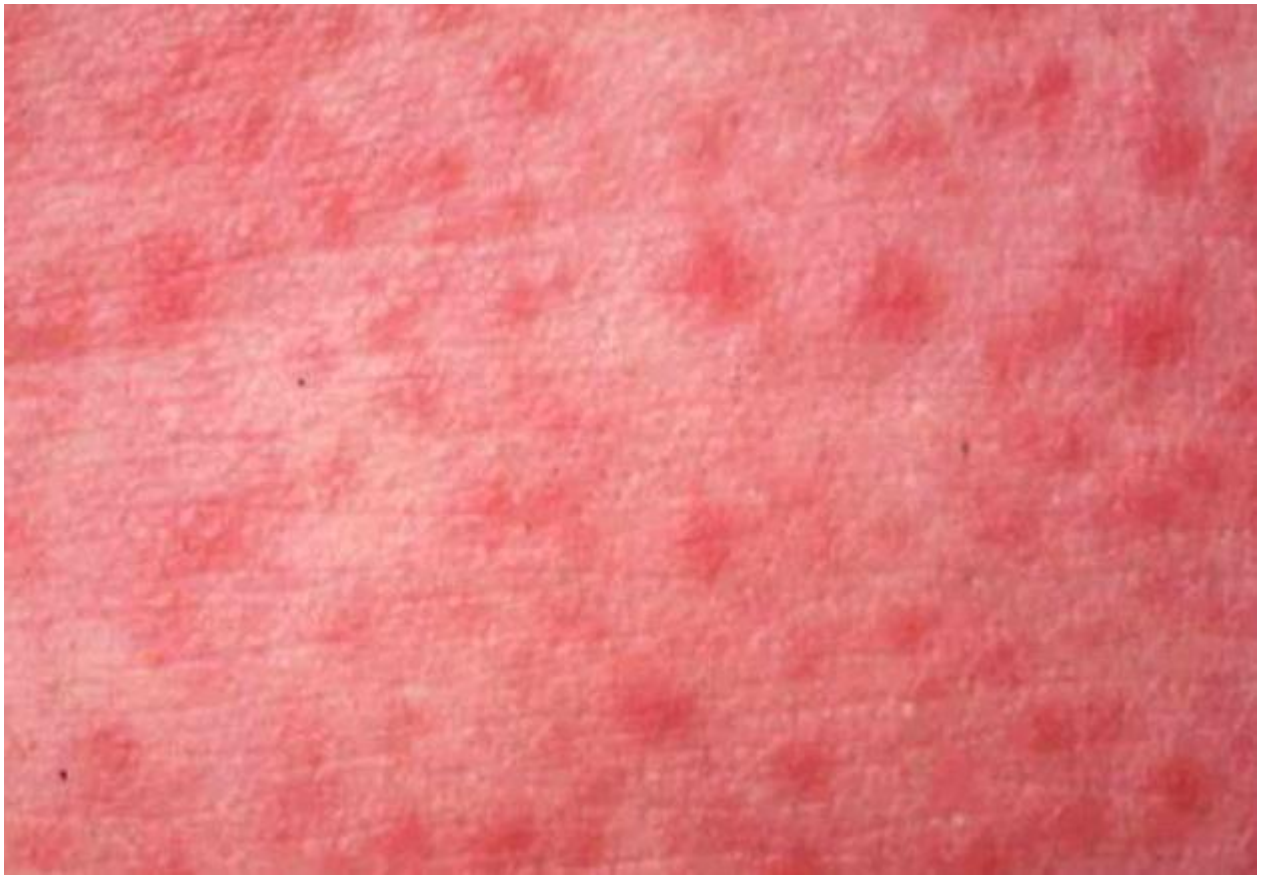
Skin disorders associated with celiac disease

Acquired ichthyosis
Cutaneous amyloid
Cutaneous vasculitis
Dermatitis herpetiformis
Eczema
Epidermal necrolysis
Nodular prurigo
Pityriasis rubra pilara
Pustular dermatitis

Erythema Herpetiformis



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Education and information

- Academy of Nutrition and Dietetics (formerly American Dietetic Association)
- (www.eatright.org)
- American Celiac Disease Alliance
- (www.americanceliac.org)
- American Gastroenterological Association
- (http://www.gastro.org/info_for_patients/2013/6/6/understanding-celiac-disease)
- Celiac Disease Foundation
- (www.celiac.org)
- Gluten Intolerance Group of North America
- (www.gluten.net)
- National Foundation for Celiac Awareness (NFCA)
- (www.celiaccentral.org)
- National Institute of Diabetes and Digestive and Kidney Diseases
- (www.niddk.nih.gov)
- National Library of Medicine
- (www.nlm.nih.gov/medlineplus/celiacdisease.html)
- North American Society for the Study of Celiac Disease
- (www.nasscd.org)
- US Food and Drug Administration
- (http://www.fda.gov/Food/GuidanceRegulation/GuidanceDocumentsRegulatoryInformation/ucm059116.htm)
- FDA and USDA labeling:
- •Thompson T. ADA Pocket Guide to Gluten-Free Strategies for Clients with Multiple Diet Restrictions, American Dietetic Association, Chicago 2011.
- •http://www.glutenfreedietitian.com/labeling-of-usda-regulated-foods

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Gluten Intolerance

- Non-celiac gluten sensitivity (NCGS) or gluten sensitivity
- **FODMAP** is an acronym, deriving from "Fermentable Oligo-, Di-, Monosaccharides And Polyols"

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