Gluten-Sensitive Enteropathy
(Coeliac Disease)

Objectives

Clinical presentations, diagnosis, D.D, complications and management.

- An autoimmune inflammatory disease of the small intestine that is precipitated by the ingestion of gluten, a component of wheat protein, in genetically susceptible persons.
- Recent studies in the United States suggest that the prevalence of celiac disease is approximately one case per 250 persons and as many as 1 in 100 UK school age children may be antibody positive for celiac disease.
- Gluten-sensitive enteropathy commonly manifests as "silent"celiac disease (i.e., minimal or no symptoms)
- Serologic tests for antibodies against endomysium, transglutaminase, and gliadin identify most patients with the disease.
- Serologic testing should be considered in patients who are at increased genetic risk for gluten-sensitive enteropathy (i.e., family history of celiac disease or personal history of type I diabetes) and in patients who have chronic diarrhea, unexplained anemia, chronic fatigue, or unexplained weight loss.

Clinical Presentation

The "classic" form typically presents in infancy at 8-24 months manifests as failure to thrive, diarrhea, abdominal distention, wasted buttock abnormal stools and irritability .

Beyond infancy, the symptoms of celiac disease tend to be less dramatic. mild non specific GI symptoms ,anemia (iron and or folate def.) and growth flattering , or is identified on screening of children at increase risk (Type 1 diabetes mellitus ,Autoimmune thyroid disease , down syndrome) and first degree relatives with known coeliac disease

Silent celiac disease

A number of investigators believe that clinically apparent gluten-sensitive enteropathy represents the "tip of the iceberg" of the overall disease burden. Patients who were detected in the seroprevalence studies were asymptomatic or oligosymptomatic (so-called "silent"celiac disease).

Diagnosis

Serologic tests

When the diagnosis of gluten-sensitive enteropathy is suspected, serologic tests can identify many affected patients.

It is important to note that 2 to 3 percent of patients with gluten-sensitive enteropathy also have selective IgA deficiency.

Antibody test	Sensitivity (%)	Specificity (%)
IgA antiendomysial antibody	85 to 100	96 to 100
IgA antitransglutaminase antibody	95	90
IgA antigliadin antibody	53 to 100	65 to 100
IgG antigliadin antibody	57 to 100	42 to 98

Distal duodenal biopsy

Is the gold standard for the diagnosis of celiac disease. Biopsy should be performed in most patients with suspected gluten-sensitive enteropathy. The characteristic features of villous flattening, crypt hyperplasia, and increased intraepithelial lymphocytes were shown to normalize after the institution of a gluten free diet.

Abnormal Laboratory Findings in Celiac Disease

Laboratory findings	Pathophysiology
*Anemia	Iron deficiency; vitamin B12 and/or folate deficiency
*Elevated alkaline phosphatase	Osteoporosis, osteomalacia
*Elevated aspartate transaminase and alanine transaminase levels	Minimal elevation common ; presumably autoimmune Malnutrition Vitamin D deficiency, secondary hyperparathyroidism
*Decreased albumin level	General inflammatory reaction
Elevated calcium level, decreased phosphate level	Decreased vitamin K absorption
Thrombocytosis, leukocytosis	Decreased fat absorption, decreased hepatic lipoprotein production
Coagulopathy	
Low high-density and low-density lipoprotein cholesterol levels	

Complications

Osteoporosis : due Calcium and vitamin D malabsorption

Neurologic manifestations

- Cerebral calcifications and epilepsy
- Peripheral neuropathy, postural instability, "gluten ataxia,"
- vague neurological complaints may be the sole manifestation
- gluten-sensitive enteropathy

Refractory sprue

In patients with refractory sprue, gastrointestinal tract inflammation continues despite maintenance of a gluten free diet. Dietary noncompliance is the most common reason for persistent inflammation;

Lymphoma and bowel adenocarcinoma

Enteropathy-associated T-cell lymphoma has been associated with untreated gluten-sensitive enteropathy and refractory sprue. Lymphoma may develop in patients with celiac disease who also have dermatitis

herpetiformis. Studies have shown that maintenance of a long term gluten-free state reduces the risk of lymphoma to the level in the general population. Patients with celiac disease are also at risk for the development of bowel adenocarcinoma in all sites.

Laboratory Evaluation of Patients with Newly Diagnosed Celiac Disease

Hematology

Complete blood cell count

Platelet count

Laboratory tests

Iron level, total iron-binding capacity determination, ferritin level*

Vitamin B12 and folate levels

Calcium and phosphate levels

Alkaline phosphatase level

Blood urea nitrogen and creatinine levels

Albumin and total serum protein levels

Aspartate transaminase and alanine transaminase levels

Imaging

Dual energy x-ray absorptiometry (DEXA) of spine and hip

Serologic tests†

Quantitative IgA antiendomysial antibody or quantitative IgAantitransglutaminase

Quantitative IgA and IgG antigliadin antibodies

Management

Once the diagnosis of celiac disease has been made, patients should be evaluated for known manifestations and complications.

- Iron deficiency should be treated with supplemental iron
- Osteoporosis should be treated with calcium and vitamin D replacement.
- Depending on individual factors, patients with gluten-sensitive enteropathy may need to take a multivitamin, iron, calcium, magnesium, zinc, selenium, vitamin D, or other nutrients.
- The primary treatment for celiac disease is the removal of gluten and related proteins from the diet.
- Complete exclusion of dietary gluten generally results in rapid and complete healing of small-bowel inflammation.

- Advice from a dietitian is essential , diet free from wheat, rye and barley result in resolution of symptoms .Meats, vegetables, fruit, and most dairy products are free of gluten, as long as they have not been contaminated during production

Key message

Although the diagnosis is strongly suggested by positive serology, confirmation depend upon the demonstration of flat mucosa on jejunal biopsy followed by the resolution of symptoms and catch growth upon gluten free diet .There is no place for empirical use of gluten -free diet as a diagnostic test for celiac disease in the absence of jejunal biopsy . Serological tests are not considered sufficiently sensitive and specific to replace biopsy as the diet is being lifelong.

Reference

Nelson text book of pediatric -web sites

Illustrated text book of pediatric