

Celiac Disease & Turner Syndrome



The Kogan Celiac Center

Topics:

What is Celiac Disease

Epidemiology of Celiac

Symptoms: Classic / Non-Classic

Associated Disorders and Risk Factors

Diet Modification

Contact Information: The Kogan Celiac Center

What is Celiac Disease?

- Celiac Disease is a permanent, genetic disorder that is characterized by an autoimmune reaction to the ingestion of gluten (a protein found in wheat, rye, and barley). This reaction causes damage to the absorption structures in the small intestines (villi) and ultimately result in:
 - Conditions of malnutrition
 - Potential damage to any organ system
- There is no cure for Celiac Disease
- The only known treatment is the complete removal of gluten from the diet.

Epidemiology

- Affects all cultures and nationalities
- 1 in 133 individuals in the U.S. are believed to have Celiac disease and it has been reported to take up to 11 years to diagnose.

The “Celiac Iceburg”



Classic Symptoms

- Anorexia
- Diarrhea
- Bloating Gas
- Vomiting
- Constipation
- Lactose intolerance
- Abdominal Pain
- Pale or foul smelling stool
- Weight Loss/Muscle Wasting

Non-Classic Symptoms

- Failure to thrive in infants
- Anemia resistant to oral iron therapy
- Autoimmune thyroid disease
- Unexplained short stature or delayed puberty
- Depression, Irritability, Learning or Behavior disorders
- Seizures
- Neuropathy/ataxia
- Recurrent fetal loss (intrauterine growth retardation)
- Delayed or missed period, infertility
- Osteoporosis/osteopenia
- Oral sores
- Loss of tooth enamel
- Vitamin Deficiencies



Associated Disorders/Risk Factors

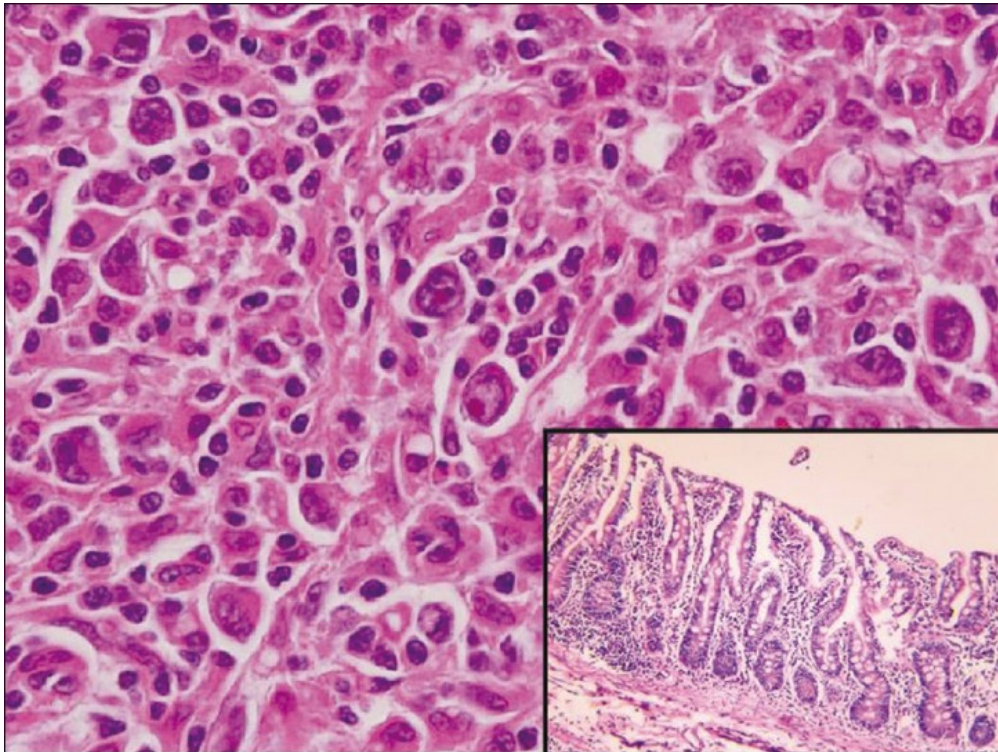
- Type 1 Diabetes
- Thyroiditis
- Addison's Disease
- Primary Biliary Cirrhosis
- Autoimmune Hepatitis
- Rheumatoid Arthritis
- Sjogren's Syndrome
- Down Syndrome
- Turner's Syndrome
- Williams Syndrome
- Increased risk of celiac disease in relatives of celiacs:
 - 7-10% primary relatives
 - 3-5% secondary relatives

Celiac Disease and Turner's Syndrome –Putting it All Together

- 2002 Italian Study
 - 25/389 (6.4%) TS patients had celiac disease
- 25 family members of the 25 also had celiac disease
- Overlap of symptoms: short stature and delayed puberty
- Before GH and estrogen therapy, screen for celiac disease

Diagnosis of Celiac Disease

- Serology
 - Total Serum IgA
 - Tissue transglutaminase antibody (tTg-IgA) (tTg-IgG if IgA deficient)
 - AGA IgA if <5 years old
- Small Bowel Biopsy
 - Villous atrophy
 - Crypt hyperplasia



Above Villous Atrophy and Crypt Hyperplasia

Dermatitis Herpetiformis

- Skin manifestation of celiac disease.
 - Itchy, pustules, bilateral
 - Dx by local skin biopsy
 - Frequently (-) serology, (+) SBB
 - Symptoms and villous atrophy respond to GFD



Above Dermatitis Herpetiformis

What is Gluten?

- Gluten is the trigger for the inflammatory response of celiac disease.
- Gluten is a general name for storage proteins
- Storage proteins in wheat, barley and rye are harmful to celiac- these grains and any ingredient that comes from them are eliminated in GFD
- Spelt, kamut and triticale are botanically related to wheat and are eliminated too.

The Gluten Free Diet

- Substitution of obvious sources- Market research shows 86% increase in new products in 2006, with continuing trend in 2007 and 2008.
 - Breads
 - Pasta
 - Baked Goods
- Consider naturally gluten free dishes
 - Risotto



- Stir-fry
- Chili
- Omelettes
- Managing unclear sources
 - Unexpected (French fries, soy sauce, rice crispies, medicine)
 - Hidden (Malt, bouillon cubes, brown rice syrup, caramel, dextrins, diglycerides, emulsifiers, fillers, flavorings, food starch, gravy, ground spices, gums, hydrolyzed plant protein, imitation bacon, imitation seafood, miso, modified starch, monoglycerides, MSG, natural color/flavor, smoke flavoring, stabilizers, textured vegetable protein, thickeners, vanilla, vegetable gum)
 - Cross contamination

The Food Allergen Labeling and Consumer Protection Act (FALCPA)

- Currently, nutrition labels must list 8 most common allergens
- Definition of gluten free claim planned for fall of 2008
- Note: Law covers dietary supplements and vitamins, NOT prescription drugs or OTC medication

The Kogan Celiac Center of the Saint Barnabas Health Care System

MISSION- To provide expert services including early assessment, diagnosis, treatment, education and support to improve the health and well being of those who live with celiac disease.

- Provides comprehensive approach to the treatment and management of celiac disease for adults and children
 - Early Assessments and diagnosis
 - Screening Services
 - Education
 - Support
 - www.koganceliacenter.com
 - 973-322-7272
 - Join Us!