

Celiac Disease

What the Clinician Needs to Know

Elaine Danner, RD, CD, CNSD
Children's Hospital of Wisconsin

September 2008

Celiac Disease

- an immune-mediated enteropathy characterized by:
 - small intestinal inflammation
 - villous atrophy
 - crypt hyperplasia
- found in **genetically** susceptible individuals
- caused by a permanent sensitivity to "gluten" in **wheat, barley and rye**
- a **multisystem** disorder so the clinical presentation is highly variable

Prevalence in General Population in U.S.

- In average healthy people: **1 in 133** (~1%)
- Celiac disease is more common in the Caucasian population – but it has been described in people of all racial and ethnic backgrounds.

Prevalence of CD

- In **first-degree relatives** of individuals with biopsy-proven CD:

4 – 12%

- Celiac disease is more common among patients diagnosed with:

■ Down syndrome	9.0-12.0%
■ Addison's disease	6.9%
■ Turner syndrome	6.4%
■ Infertility (idiopathic)	6.2%
■ Type 1 diabetes mellitus	4.3% - 10.3%
■ Autoimmune thyroiditis	4.3%
■ Anemia	4.2%
■ Short stature	4.0%
■ Chronic diarrhea	3.8%
■ Abdominal pain	3.2%
■ Arthritis	3.0%
■ Primary biliary cirrhosis	3.0%
■ Fatigue	2.9%
■ Constipation	2.6%
■ Asthma	2.6%
■ Osteoporosis	2.6%
■ Sjogren syndrome	2.0-5.0%

Facciano A, 2003
Ashabani A, 2003
Myhre AG, 2003
Bonamico M, 2002

U.S. National Institutes of Health - 2004

Six Key Elements In The Treatment Of Celiac Disease

- **Consultation** with a skilled dietician
- **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**

How is Celiac Disease Diagnosed?

- There is no one test that can definitively diagnose or exclude celiac disease.
- The combination of clinical symptoms and laboratory results may result in a diagnosis of CD

Clinical Symptoms

- Gastrointestinal – if any, may include:
 - Diarrhea
 - Weight loss*
 - Failure to grow
 - Vomiting
 - Abdominal pain
 - Bloating and distention
 - Anorexia
 - Constipation
- *Obesity does not exclude the diagnosis of CD

Clinical Symptoms

- Non-gastrointestinal – if any, may include:
 - Dermatitis Herpetiformis – an intensely pruritic rash
 - Iron deficiency anemia
 - Short stature
 - Delayed puberty
 - Infertility and/or recurrent miscarriages
 - Osteoporosis
 - Vitamin deficiencies
 - Fatigue
 - Protein caloric malnutrition
 - Recurrent aphthous stomatitis (canker sores)
 - Dental enamel hypoplasia

Clinical Symptoms

- CD may also be associated with:
 - Autoimmune endocrinologic disorders such as thyroiditis
 - Neuropsychiatric conditions such as depression, anxiety, peripheral neuropathy, ataxia, epilepsy with or without cerebral calcifications and migraine headaches.

Laboratory Results

- Important to note **all diagnostic tests need to be performed while the patient is on a gluten-containing diet**
- Only **exception** is the genetic testing for HLA haplotypes

Laboratory Tests – Serology

- **Best available tests:**
 - IgA antihuman tissue transglutaminase (tTG-IgA)
 - IgA endomysial antibody immunofluorescence (EMA-IgA)
- A **total IgA** is usually done in conjunction with these tests to rule out a selective IgA deficiency
- If a patient is **IgA deficient** then a tTG-IgG or an EMA-IgG can be ordered
- **Antigliadin antibody (AGA)** tests are **not recommended** because of their lower sensitivity and specificity
- Serologic testing for CD in children younger than 5 years of age **may not** be reliable

EGD with Biopsies – Histology

- **Multiple biopsies** should be obtained since the histologic changes may be focal
- Biopsies should be obtained from the second portion of the duodenum or beyond
- The pathology report should specify:
 - the degree of crypt hyperplasia and villous atrophy
 - the number of intraepithelial lymphocytes

Genetic Testing

- A means of **ruling out** CD
- If a **diagnosis** of CD is **uncertain** based on serology and/or histology – testing for certain genetic markers can be done
- >97% of CD individuals have the HLA-DQ2 and/or DQ8 marker
- An individual **negative** for HLA-DQ2 or DQ8 is **extremely unlikely** to have CD

Gastrointestinal – “Classic” Celiac Disease

- Most common age of presentation: 6-24 months
- Symptoms begin within few months of introducing foods containing prolamins
- Characterized by:
 - Chronic, recurrent diarrhea
 - Abdominal pain
 - Abdominal distension
 - Vomiting
 - FTT or weight loss
 - Constipation
 - Irritability
 - Anorexia

Fasano 2001

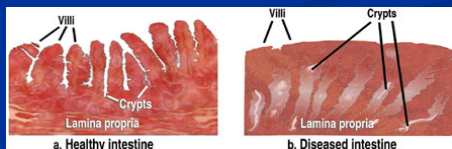
Examples of Presentations of Celiac Disease

- College student with frequent bruising
- Pre-menopausal woman with osteoporosis
- 82 year-old admitted with weight loss and malnutrition
- 40 year-old male with iron deficiency anemia not amenable to oral iron-supplementation

Other Possible Intolerances

Disaccharidase (enzymes that break down sugars) production can be affected.

Lactose-intolerance that results will likely be temporary until the damage to the villi is resolved.



Gluten

- general name for storage proteins called **prolamins** that are found in various cereal grains.
- The specific names of these prolamins are:
 - **Gliadin** in wheat
 - **Secalin** in rye
 - **Hordein** in barley
- It is the specific amino acid sequence (possibly a 33 amino acid molecule) in these prolamins that triggers the immune response in CD.

Oats

- Currently the inclusion of oats in the GF diet remains an area of **controversy**.
- The prolamin in oats is avenin

The Key Points of Controversy

- Research regarding the safety of oats for people with celiac disease is still evolving
- Gluten-contamination of oat products

Research

- Numerous studies have shown that “research grade” oats are safe for celiacs.
- Included is a 5-year F/U study done by Janatuinen and colleagues that looked at the long-term effect of including oats every day (mean intake of 34 g/day) – **concluded that long-term consumption of a moderate amount of oats by adults with CD was safe**

Kemppainen 2007
Janatuinen 2002

Gluten-contamination Of Oat Products

Contamination may be due to or occur during:

- Planting with seed contamination
- Crop rotation
- Harvesting equipment
- Transportation
- Processing Facility

Gluten –Free Oats

- Gluten Free Oats®
 - www.glutenfreeoats.com
 - Per website: tested to be below 10 parts per million (ppm)
- Cream Hill Estates
 - www.creamhillestates.com
 - Per website: Our oats consistently test below or near the test's lower limit of detection, which is two (2) parts per million.

Grains to Avoid

- Wheat (durum, semolina, einkorn, emmer, farro, graham, wheat bran, wheat starch, wheat germ)
- Rye
- Barley
- Triticale – a hybrid of wheat and rye
- Kamut – a form of wheat
- **Spelt** – a form of wheat
- Bulgur – a form of whole wheat

What Foods Contain Barley??

- Barley Soup
- Beer and Malt Beverages such as Smirnoff Ice™, Mike's Hard Lemonade™, Bartles & Jaymes™, etc.
- Barley Malt Flavoring
 - Malted shakes
 - Many cereals such as Rice Krispies™, etc.
 - Many other foods (can be listed as a "natural" flavoring)
- Barley Extract
 - Found in some rice and soy milks
 - Some chocolate milks

What Foods Contain Rye??

- Mainly rye-flavored breads and crackers
- Usually a secondary ingredient and will be included in the list of ingredients
- Use of rye extract in meat products

What Foods Can Contain Wheat??

Breads/Rolls/Buns/Bagels	Biscuits
Crackers/Matzo	Dumplings
Cakes and Cookies	Doughnuts
Pasta/Noodles	Pretzels
Pizza	Pie Crust
Cereals	Stuffing
Pancakes and Waffles	Breaded foods

What Other Foods Can Contain Wheat??

- Ice cream cones
- Tortillas and wraps
- Couscous
- Soy and Worcestershire sauces
- Gravies
- Cream sauces
- Seasoning mixes
- Broth/Bouillon/Soup Bases/Canned Soups
- Candies
- Communion wafers
- Marinades
- Dry roasted nuts
- Icings and frostings
- Imitation seafood and meats
- Vegetarian "meat" substitutes
- Thickeners, stabilizers, texture enhancers

Communion Wafers

- Gluten-free wafers are available
- Catholic Canon Law states that the wafer must be made from wheat and water
- A low-gluten wafer has been developed
- Carlo Catassi and Alessio Fasano (U of MD) have indicated that the low-gluten wafers are safe/allowable
- Other recommendations:
 - take only a piece of the wafer
 - use a **pyx** to prevent contamination

Non-Food Sources of Gluten

- Medications – both prescription and non-prescription
- Play-Doh™
- Toothpaste
- Lip balm, lip gloss and lipstick
- Stamps, envelopes
- Stickers
- Pet food and animal feeds
- Air-borne gluten

Other Sources of Gluten

- Contamination
 - Toaster
 - Butter dish, margarine container, peanut butter and jelly jars
 - Deep-fat fryers
 - Conveyor belts
 - Bins and scoops
 - Deli slicer
 - Starch coating on wrappers to prevent sticking

Allowable Grains, Seeds and Starches

- Amaranth*
- Arrowroot
- Buckwheat*
- Corn
- Legumes*
- Montina*
- Millet
- Nuts*
- Potato
- Quinoa*
- Rice
- Sorghum/Milo*
- Soy*
- Tapioca
- Tef/Teff*
- Wild Rice

* Tend to have more nutrient(s) i.e. more fiber, protein, calcium, iron

Food Allergen Labeling and Consumer Protection Act FALCPA

- took effect January 1, 2006
- mandates that foods containing one of the "top 8 allergens" must have the allergen declared in plain language on the label.

Top 8 Allergens	
Milk	Eggs
Peanuts	Tree Nuts
Crustacean Shellfish	Fish
Soybeans	Wheat

FALCPA

- applies to both domestically manufactured and imported packaged foods that are subject to FDA regulation
- includes flavorings, colorings, and incidental additives

FALCPA

- Rye, Barley and Oats are **not** listed as allergens
- requires FDA to issue a final rule that will define and permit the voluntary use of the term "**gluten free**" on the labeling of foods by August 2008.

Current Draft of Proposed Rule

- Gluten-free is defined as **less than 20 ppm** gluten in food
- **Gluten-free** for labeling means that foods **do not contain** any of the following:
 - An ingredient that contain **any species of the grains** wheat, rye, barley, or a crossbred hybrid of these grains (known collectively as "prohibitive grains")
 - An ingredient that is derived from a prohibitive grain and **has not been processed** to remove the gluten (such as wheat flour)
 - An ingredient that is derived from a prohibitive grain and **has been processed** to remove the gluten (such as wheat starch), **and** if the use of that ingredient results in the food having 20 ppm or more gluten.
- **20 ppm or more** gluten in the food.

How much gluten is too much??

- Hischenhuber and colleagues reviewed a number of studies and concluded that the maximum tolerated daily intake of gluten is > 10 mg and < 100 mg of gluten
- This equates to 1/50th – 1/500th of a slice of bread
- Catassi and colleagues in a double-blind, placebo-controlled, randomized trial in 49 adults concluded that the ingestion of gluten should be <50 mg/day.

Hischenhuber 2006
Catassi 2007

Nutritional Deficiencies

At the time of diagnosis

Might include:

- Calories/Protein
- Iron
- Calcium
- Fat-soluble vitamins
- Zinc
- Folate
- B₁₂

Nutritional Deficiencies

Related to Lack of Enrichment/Fortification
of GF Products

Might include:

- Fiber
- Iron
- Folate, niacin, thiamin, riboflavin

Medications

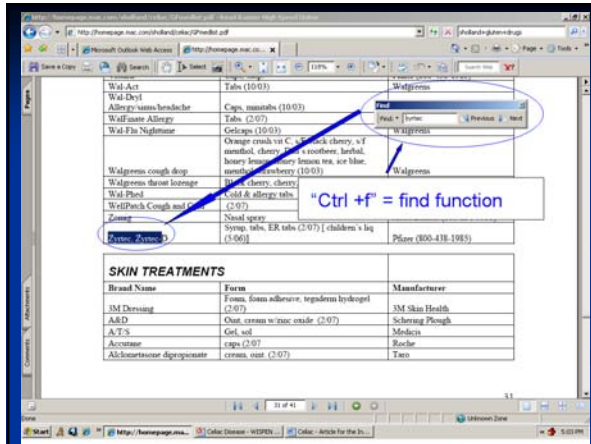
- Gluten in medications comes mainly from the excipient (filler) ingredients
- Some ingredients that may contain gluten:
 - Starch, pregelatinized starch, sodium starch glycolate
 - Dextrates and dextrans
- Generic product does not have to contain the same excipients as the brand name product
- Check frequently with the pharmaceutical company as ingredients may change

Medications

- The federal government does not place as many controls on over-the-counter drugs as they do with prescription drugs
- Nutritional supplements fall under the Food Allergen Labeling and Consumer Protection Act of 2004

Gluten-Free Pharmaceutical Resources

- Printed References – including package inserts
- Manufacturer's Customer Service or Website
- Pharmacist
- www.glutenfreedrugs.com
- <http://homepage.mac.com/sholland/ceeliac/GFmedlist.pdf>




Celiac Diet Series Articles from the journal *Practical Gastroenterology*

- <http://www.healthsystem.virginia.edu/internet/digestive-health/nutrition/celiacsupport.cfm>
- Series of electronic articles including:
 - Medications and Celiac Disease – Tips From a Pharmacist*
 - The Gluten-Free Diet: An Update for Health Professionals*
 - Combining Diabetes and Gluten-Free Dietary Management Guidelines*
 - The Gluten Free Vegetarian*
 - Others

Celiac Sprue Association

www.csaceliacs.org

CSA Product Listing Guide



- Published annually in October by the Celiac Sprue Association
- Contents include listings of GF products according to info obtained from manufacturers + distributors.
- Phone numbers, etc. of manufacturers
- Extensive glossary

Gluten Intolerance Group

www.gluten.net

The following GIG publications may be **downloaded** from this website **free of charge**.

- [Quick Start Diet Guide](#)
- [Adding Fiber to Your Gluten-Free Diet](#)
- [Gluten Sensitivity](#)
- [Celiac Disease](#)
- [Dermatitis, Herpetiformis](#)
- [Associated Autoimmune Diseases](#)
- [Diabetes and Celiac Disease](#)
- [Understanding Your Student](#)
- [Restaurant Dining Info](#)
- [Travel in the US](#)
- [About GIG®](#)
- [Celiac, Diabetes, and Me!](#)
- [Easy-to-Find and Fix Foods](#)
- Others

Clan Thompson Celiac

<http://www.clanthompson.com/index.php3>

- Smart List Database or Booklets can be purchased
- Free Information:
 - Electronic Newsletter
 - Gluten Alerts
 - Free Lists

Frito Lay

www.fritolay.com

Wendy's

www.wendys.com

Impact on Food Buying

- Gluten-free diet tends to include more meats, fruits and vegetables - which are naturally gluten-free and readily available but ↑ cost
- Availability of packaged gluten-free products may be limited (scout out your stores especially local grocery stores)
- Gluten-free products may cost 2 – 3 times as much

Lee 2007

edanner@chw.org

Include "*Celiac References*" in subject line