Celiac Disease: Beaten by the Staff of Life

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Celiac Disease: A Definition

- Auto Immune Disease
- Triggered by Environmental Exposure to Gluten
- Genetically Susceptible Individual
- Principally an Enteropathy
- Multisystem Effects
Celiac Disease: Statistics in the United States

- In average healthy people: 1 in 133
- In people with related symptoms: 1 in 56
- In people with first-degree relatives
  (parent, child, sibling) who are celiac: 1 in 22
- In people with second-degree relatives
  (aunt, uncle, cousin) who are celiac: 1 in 39
- Estimated prevalence for African-

Celiac Disease: A History

- Man evolved from earlier hominids as a hunter/gather over approximately 2,000,000 years
  - Gut with active immune system evolved to cope with presentation of variety or nutrients containing foreign proteins
  - Diet consisted mainly of fruits, nuts, tubers, insects, eggs, and occasional meat
Celiac Disease: A History

• Agriculture developed about 10,000 years ago, adding new exposures to the gut
  – Dairy Products: Milk from Cows, Sheep/Goats/Donkeys
  – Vegetables
  – Grains

• First Described in Greek Times
  – 1st Century CE
  – Aretaeus of Cappadocia, a Greek Physician
  – Wrote about “The Coeliac Affliction”
  – Described symptoms of malabsorption and maldigestion
Celiac Disease: A History

• Early 19th Century
• Matthew Baillie, M.D. (1761-1823)
  – Described a chronic diarrheal disease causing a distended, gas filled abdomen and malnutrition
  – Noted that symptoms seemed to improve on a diet consisting mainly of rice.
  – Went Largely Unnoticed

• Late 19th Century
• Dr. Samuel Gee (1839-1911)
  – Expert in Pediatric Disease
  – Credited with Modern Description of Celiac Disease.
  – Taught Medical Students about “The Coeliac Affliction
Celiac Disease: A History

• Dr Samuel Gee (1839-1911)
  – Made Key Observations of his patients
    • Described an affected Dutch Child
      – Was healthy Eating 1 Quart Mussels Daily
      – Became ill after Mussel Season Ended
      – Improved after withdrawal of “Farinaceous Foods” from Diet
      – Relapsed with their reintroduction

Celiac Disease: A History

• Early 20th Century: Sidney Haas (1870-1964)
  – 1920s: The Banana Diet
    • 10 Children with Anorexia, Diagnosed with Celiac Disease
      – 8 were Treated described as “Cured”
      – 2 Untreated, all died
  – Excluded Carbohydrates
    • Bread, Potatoes, and all Cereal Grains
  – Became the Mainstay of Treatment for Decades
Celiac Disease: A History

• Willem-Karel Dicke (1905-1962)
  – Dutch Physician
  – Made Observations During World War II
    • Chronic Bread Shortages Secondary to Farmland Destruction
    • Everyone losing weight except certain Children who seemed to thrive
    • The Children would worsen quickly after Allied Planes dropped foods, especially bread into Cities

Celiac Disease: A History

• Margot Shiner, M.D. (1923-1998)
• Pediatric Pathologist and Gastroenterologist
• Developed Small Bowel Biopsy Capsule (1956)
  – Allowed Non Surgical Biopsy of the Small Intestine
  – Applied Jejunal Biopsy Technique to Duodenum
• William H. Crosby, Jr., M.D. (1914-2005)
  – Refined the Biopsy Capsule
Celiac Disease: Diagnosis
Clinical Presentation

• More than 200 associated Signs and Symptoms:

  Recurrent Abdominal Pain/Bloating  Iron Deficiency
  Chronic Diarrhea/Constipation  Peripheral Neuropathy
  Vomiting  Osteopenia/Osteoporosis
  Weight Loss  Failure to Thrive
  Pale, Foul-Smelling Stool  Short Stature
  Joint Pain  Delayed Puberty
  Dermatitis Herpetiformis
  Unexplained Infertility

Celiac Disease: Diagnosis
Clinical Presentation

• Many Have No Symptoms!
  – The undamaged portion of the small intestine is
    sufficient to absorb sufficient nutrients to prevent
    development of symptoms

• Classic Presentation: Malabsorption and Maldigestion
  – Diarrhea/Steatorrhea/Iron Deficiency Anemia
Celiac Disease: Diagnosis
Clinical Presentation

• In Children
  – Failure to Thrive
  – Growth Retardation
  – Abdominal Pain and Bloating
  – Diarrhea/Constipation
  – Fatigue Irritability

Celiac Disease: Diagnosis

• 1969: European Society for Pediatric Gastroenterology: “The Interlaken Criteria”
  – Mucosal Atrophy on Small Bowel Biopsy
  – Clinical Remission on Gluten-Free Diet
  – Reversal of Mucosal Abnormalities on Gluten-Free Diet
Celiac Disease: Diagnosis

• Mechanism for Celiac Disease
  – Anti-Gliadin Antibody in Children with Celiac Disease
    • An Anti Food Antibody
    • First Described by E. Berger in 1964
  – Anti Reticulin Antibody 1971
    • First Anti Tissue Antibody
    • Connects Immune Reaction to Food Protein to Immune Mediated Tissue Damage

Celiac Disease: Diagnosis

• More Recent Developments
  – Specific Gene Associations
    • DQ2 or DQ8
• Leading to the conclusion:
  – Celiac Disease is an autoimmune disease in which Gluten is the Trigger and Tissue Transglutaminase is the Autoantigen
Celiac Disease: Testing Modalities

• Clinical Suspicion Leads to Testing
• Serologic Tests: A good initial screen
  – Most are IgA Antibodies
    • Check total serum IgA level; if IgA level low, get IgG Antibodies
  – Antigliadin Antibody (AGA)
  – Anti Endomysial Antibodies (EMA)
  – Anti Tissue Transgluaminase Antibody (tTG)
• Mucosal Biopsy
  – The Gold Standard

Celiac Disease: Testing Modalities

• tTG Testing
  – ELISA assay
  – >90% Sensitivity
  – >95% Specificity
• EMA
  – Less sensitivity than tTG, but near 100% Specificity
• AGA
  – Neither sensitive nor specific to use routinely except in children younger than 18 months
Celiac Disease: Testing Modalities

Genetic Testing

- HLA Variants
  - DQ2
  - DQ8
- Above are common
  - 40% of US Population
  - Most do not have Celiac disease

Celiac Disease: Treatment Modalities

- Lifelong Elimination of Gluten from the Diet
- What is Gluten?
  - Grain Storage Proteins
    - Gliadin and Glutenin in Wheat
    - Hordein in Barley
    - Secalin in Rye
    - Avenin in Oats
    - Zein in Corn
    - Oryzenin in Rice
  - Corn and Rice gluten do not trigger Celiac Disease
  - Contain Amino Acid Sequences triggering Celiac Disease.
Celiac Disease: Treatment
A Gluten Free Diet

• Grains which are OK
  – Rice, Corn, Soy, Potato, Tapioca, Beans, Quinoa, Buckwheat, Arrowroot, Nut Flours, Garfava, Montina, Amaranth

• Grains which will Harm
  – Wheat (Includes semolina, durum, graham, kamut, spelt), Rye, Barley, Tricale, Farina

Celiac Disease: Treatment
A Gluten Free Diet

• Reading Labels: A Key to Success with Celiac Disease. Avoid Foods containing:
  – Malt or Malt Flavoring
  – Malt Vinegar
  – Textured Vegetable Protein
  – Any of the prohibited Grains
Celiac Disease: Treatment
A Gluten Free Diet

• Foods to Avoid:
  – candy bars
  – canned soup
  – canned meat
  – energy bars
  – Ketchup
  – ice cream
  – instant coffee
  – lunch meat
  – Mustard
  – Pastas
  – processed foods

Celiac Disease: Treatment
Other Advice

• Alcohol
  – Avoid Beer
  – Whiskey/Spirits, Wine, Brandy all fine: gluten does not pass through distillation

• Cosmetics may use wheat starch as a binder
• Check with pharmacist regarding pills and capsules
• Cookware/utensils
Celiac Disease: Complications

- Malnutrition
- Bone and Mineral Density Loss
- Lactose/Fructose maldigestion
- Neurologic Issues
  - Increased risk of seizure disorder
  - Peripheral Neuropathy
- Increased Risk of Cancer
  - Colon Cancer
  - Small Bowel Lymphoma
  - Small
  - Decrease with Maintaince of Celiac Diet

Celiac Disease Follow Up

- Repeat Serologic Testing
  - tTG IgA and AGA IgA and IgG
    - Aim to see them as close to zero as possible
  - 3-6 months after diagnosis
  - 1 yr following diagnosis and yearly thereafter
Celiac Disease: The Future

- Altered Grain: genetic engineering
- Immunomodulatory Therapy
- Enzyme Therapy
- Repair Intestinal Permiability Defect