Diagnosis and Management of Children and Adolescents with Celiac Disease

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Celiac disease is an immune-mediated enteropathy caused by a permanent sensitivity to gluten in genetically susceptible individuals.
Clinical Manifestations

- Gastrointestinal ("classical")
- Non-gastrointestinal ("atypical")
- Asymptomatic

In addition, Celiac Disease may be associated with other conditions, and mostly with:
- Autoimmune disorders
- Some syndromes
The Celiac Iceberg

- Symptomatic Celiac Disease
- Silent Celiac Disease
- Latent Celiac Disease

Genetic susceptibility: - DQ2, DQ8
Positive serology

Manifest mucosal lesion
Normal Mucosa
Gastrointestinal Manifestations ("Classic")

Most common age of presentation: 6-24 months

- Chronic or recurrent diarrhea
- Abdominal distension
- Anorexia
- Failure to thrive or weight loss

Rarely: Celiac crisis

- Abdominal pain
- Vomiting
- Constipation
- Irritability
Typical Celiac Disease
Non Gastrointestinal Manifestations

Most common age of presentation: older child to adult

- Dermatitis Herpetiformis
- Dental enamel hypoplasia of permanent teeth
- Osteopenia/Osteoporosis
- Short Stature
- Delayed Puberty

- Iron-deficient anemia resistant to oral Fe
- Hepatitis
- Arthritis
- Epilepsy with occipital calcifications

Listed in descending order of strength of evidence
Dermatitis Herpetiformis

- Erythematous macule > urticarial papule > tense vesicles
- Severe pruritus
- Symmetric distribution
- 90% no GI symptoms
- 75% villous atrophy
- Gluten sensitive

Dental Enamel Defects

Involve the secondary dentition
May be the only presenting sign of Celiac Disease
Osteoporosis

Low bone mineral density improves in children on a gluten-free diet.
Recurrent Aphtous Stomatitis

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Short Stature/Delayed Puberty

- Short stature in children / teens:
  - ~10% of short children and teens have evidence of celiac disease
- Delayed menarche:
  - Higher prevalence in teens with untreated Celiac Disease
Fe-Deficient Anemia Resistant to Oral Fe

- Most common non-GI manifestation in some adult studies
- 5-8% of adults with unexplained iron deficiency anemia have Celiac Disease
- In children with newly diagnosed Celiac Disease:
  - Anemia is common
  - Little evidence that Celiac Disease is common in children presenting with anemia
Associated Conditions

The prevalence of Celiac Disease is higher in patients who have the following:

- Certain genetic disorders or syndromes
- Other autoimmune conditions
- Relative of a biopsy-proven celiac
Associated Conditions

Percentage

- Relatives
- IDDM
- Thyroiditis
- Down syndrome

General Population
“Mines” of Celiac Disease Were Found Among:

- Relatives
  - short stature, anaemia, fatigue, hypertransaminasemia
- Patients with Associated diseases
  - autoimmune disorders, Down's, IgA deficiency, neuropathies, osteoporosis, infertility
- “Healthy” groups
  - blood donors, students, general population
Genetics

• Strong HLA association
• 90 - 95% of patients HLA-DQ2 – also found in 20 - 30% of controls
  – Most of the remainder are HLA - DQ8
Diagnostic principles

• Confirm diagnosis before treating
  – Diagnosis of Celiac Disease mandates a strict gluten-free diet for life
    • following the diet is not easy
    • QOL implications

• Failure to treat has potential long term adverse health consequences
  • increased morbidity and mortality
Serological Tests

Role of serological tests:

• Identify symptomatic individuals who need a biopsy
• Screening of asymptomatic “at risk” individuals
• Supportive evidence for the diagnosis
• Monitoring dietary compliance
Endoscopic Findings

Normal Appearing

Scallopning

Nodularity
Biopsy Diagnosis

- Histologic Features:
  - Increased IEL’s ( > 30/100 enterocytes)
  - Loss of nuclear polarity
  - Change from columnar to cuboid
  - Lamina propria cellular infiltrate
  - Crypt elongation and hyperplasia
  - Increased crypt mitotic index
  - Progressive villous flattening
Fantastic Voyage

Celiac

Normal
Treatment

• Only treatment for celiac disease is a gluten-free diet (GFD)
  – Strict, lifelong diet
  – Avoid:
    • Wheat
    • Rye
    • Barley
Sources of Gluten

• OBVIOUS SOURCES
  – Bread
  – Bagels
  – Cakes
  – Cereal
  – Cookies
  – Pasta / noodles
  – Pastries / pies
  – Rolls
Sources of Gluten

- POTENTIAL SOURCES
  - Candy
  - Communion wafers
  - Cured Pork Products
  - Drink mixes
  - Gravy
  - Imitation meat / seafood
  - Sauce
  - Self-basting turkeys
  - Soy sauce
Other Items to Consider

- Lipstick/Gloss/Balms
- Mouthwash/Toothpaste
- Play Dough
- Stamp and Envelope Glues
- Vitamin, Herbal, and Mineral preparations
- Prescription or OTC Medications
Recent Survey

- 78% of married women with celiac disease believe their husbands contain gluten, and should therefore be strictly avoided.
Lactose Intolerance & Celiac Disease: Incidence

- Secondary lactase deficiency is estimated to be 20-40%
- Increasing lactose Intolerance with delayed diagnosis
- Increased incidence in patients with GI symptoms in Celiac Disease
- Decrease calcium and vitamin D intake in lactose intolerance