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Celiac Disease

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Introduction

While seeming like a recent discovery, celiac disease was believed to be first described around the First Century A.D. by the Greek physician Arataeus, who referred to celiac disease as an intestinal disorder associated with malabsorption and diarrhea (Sollid & Jabri, 2013). In the 1940s, Dutch pediatrician William-Karel Dicke, discovered that celiac disease was caused by the consumption of cereal gluten proteins (Sollid & Jabri, 2013). Today scientists and healthcare providers have a much larger knowledge base on celiac disease. However, due to the considerable symptom variance among patients, celiac disease may still go undiagnosed or be mistaken for another disease process.

- Patients with celiac disease may wait as long as 11 years before a correct diagnosis is made (Reinhardt & Yancon, 2012).
- About one-third of patients have been previously diagnosed with irritable bowel syndrome (Reinhardt & Yancon, 2012).
- Celiac disease has become the most common autoimmune inherited disorder (Bacigalupe & Plocha, 2015).
- Approximately 1% of the U.S. population is affected by celiac disease (Bacigalupe & Plocha, 2015).
- Non-Hispanic whites have the highest incidence rate of 1%, signifying that at least 1.7 million in this population have celiac disease, compared to the national average of 0.71% (Mistler, 2015).
- Disease onset typically occurs between 6 months to 2 years of age after gluten has been introduced, or between 20 to 40 years of age (Reinhardt & Yancon, 2012).
- Celiac disease can range from asymptomatic to malabsorption in severe cases.

Celiac disease occurs in genetically predisposed individuals, and is triggered by the ingestion of gluten (Bacigalupe & Plocha, 2015). Gluten is a protein found in wheat, barley, and rye products. Ingestion of gluten triggers an immune reaction, which causes damage to the small intestinal mucosa (Walter, 2013). Intestinal villi, the fingerlike projections of the small bowel mucosa become partially or completely atrophied from gluten ingestion in these genetically predisposed individuals, which leads to malabsorption, possible weight loss, and risk of malnutrition (Sollid & Jabri, 2013). The only treatment for celiac disease is a life-long gluten free diet, which will allow intestinal mucosa to heal over time. Diagnostic serology must be completed before gluten is eliminated from the diet to obtain accurate results.

As a future family nurse practitioner, digestive disorders are among a long list of conditions patients may seek care for. In recent years celiac disease is a digestive disorder that has received much attention from both patients and the healthcare community. Only through proper education will healthcare professionals be able to understand common symptoms, pathophysiology, and diagnostic tools of celiac disease, which will in turn allow patients to be correctly instructed and diagnosed in a timely manner.

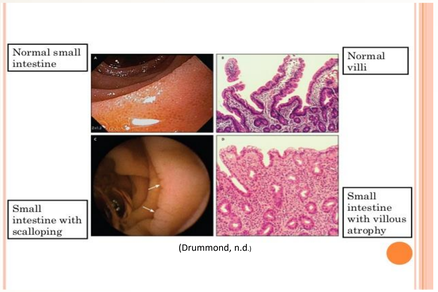
Pathophysiology

- Celiac disease is a digestive disorder that occurs when an individual's immune system overreacts to the protein gluten (Ulbricht, 2013).
- Wheat, rye, and barley glutens are the main offenders of the disease (Mistler, 2015).
- Villous atrophy is the hallmark sign of celiac disease.
- The exact mechanisms of the disease process are still being studied.

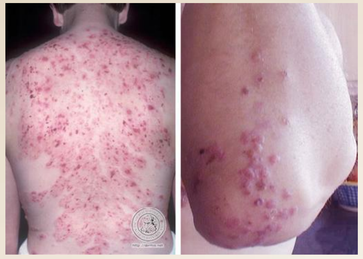
According to Mistler (2015), "proline- and glutamine-rich residues compose the protein subunits of gliadins and glutenins which make up the wheat, rye and barley glutens" that are the main offenders of the disease (p. 213). This pathophysiology is significant, as a large portion of most diets consist of grains, especially wheat. An increased amount of interferon-γ (INFγ) is secreted by CD4+ αβ T cells, coded for by the HLA-DQ2 variant 2.5 and HLA-DQ8 genes, as they become sensitized to these gluten proteins after translation modification deamidation by transglutaminase (Mistler, 2015). HLA-DQ2 and HLA-DQ8 molecules are only responsible for approximately 40% of genetic predisposing factors in the pathogenesis of celiac disease, which is necessary but not sufficient to cause disease, and therefore "many more risk loci outside the HLA region should be identified as disease markers" (Huang et al., 2017, p. 2).

Transglutaminase is the primary autoantigen seen in celiac disease, and is an enzyme responsible for transamination, or cross-linking, and deamination of glutamine residues, such as gluten (Mistler, 2015). Villous atrophy, is the "histomorphologic hallmark of celiac disease" (Shalimar, Das, Sreenivas, Gupta, Panda & Makharia, 2013, p. 1262). Villous atrophy occurs due to sensitization, allowing T cell infiltration of the mucosa. This leads to damage, villous atrophy, crypt hyperplasia and eventual activation of B cells which produce IgA antibodies to gliadin, endomysium and transglutaminase (Mistler, 2015).

The precise trigger of this reaction has not been determined, however, it is associated with autoimmune disorders, such as systemic lupus erythematosus (Ulbricht, 2013). Autoimmune disorders occur when a patient's immune system mistakenly identifies body cells as harmful invaders and as a result, the immune cells in a patient with celiac disease attack that patient's intestinal cells (Ulbricht, 2013). According to Shalimar et al., (2013), villous atrophy could occur due to increased cell destruction or enhanced apoptosis and/or a defect in epithelial cell regeneration. Damage to intestinal villi can then lead to malabsorption and malnutrition in severe cases.



(Drummond, n.d.)



Dermatitis herpetiformis caused by celiac disease. (Dermatitis herpetiformis, 2014)

Signs and Symptoms

Celiac disease can present with a wide array of signs and symptoms and may be affected by the patients age and other illnesses. Patients with celiac disease may present with various vitamin and mineral deficiencies due to intestinal damage. The most common deficiencies are vitamin D, calcium, folate, vitamin B12 and iron (Jurgelewicz, 2015). While celiac disease may result in a wide range of complications the most serious are failure to thrive and even lymphoma if celiac disease goes untreated or undiscovered long enough. According to Mistler (2015), there are four main presentations of celiac disease that have been determined by a task force of physicians from seven different countries: 1) classic, 2) non-classic, 3) subclinical, and 4) potential.

Classic

- Pediatric patients may present with: chronic diarrhea, vomiting, anorexia, abdominal pain and distention, decreased weight gain, weight loss, or nutritional deficiency (Mistler, 2015).
- Nutritional deficiency in the pediatric population, can lead to growth and psychomotor delays, rickets, and other hematological symptoms, while infants may present with severe diarrhea, dehydration, lethargy, and marked abdominal distention, which left untreated or undiagnosed can cause severe malnutrition (Mistler, 2015).
- Adults may present with constipation, nausea, loss of appetite, or intermittent diarrhea (Mistler, 2015).

Non-classic

- "Non-classic celiac disease does not manifest with typical gastrointestinal distress and may account for as much as 70% of cases" (Mistler, 2015, p. 213).
- Most of the symptoms are due to the nutritional deficiency that results from malabsorption in the small intestines due to villous atrophy, seen in the majority of pediatric cases, or in anemia in the teenage and young adult population (Mistler, 2015).
- Intestinal manifestations are usually mild gastrointestinal distress.
- Extra-intestinal manifestations include: anemia, height and weight deficiency, failure to thrive, delayed puberty, dermatitis herpetiformis, dental enamel hypoplasia, iron deficiency anemia resistant to oral therapy, low bone mineral density, liver dysfunction, neurologic disorders of depression, anxiety, autism, peripheral neuropathy, cerebral ataxia, epilepsy and migraines (Mistler, 2015).

Subclinical

- In subclinical celiac disease, many patients already suffer from another autoimmune disease such as diabetes type 1, thyroiditis, psoriasis, or genetic conditions such as Downs Syndrome, Turners Syndrome, and IgA deficiency (Mistler, 2015).
- Subclinical celiac disease is rare and patients usually do not display any signs or symptoms.

Potential

- Patients may exhibit constipation, nausea, loss of appetite, or intermittent diarrhea, but serology and biopsy results are negative for celiac disease (Mistler, 2015).

Diagnosis

- The serological tests used for detecting celiac disease include: detection of anti-tTG antibodies (a-tTG), antiendomysial antibodies (EMA), and antigliadin antibodies (AGA) (Porcelli, Ferretti, Vindigni, & Terzuoli, 2016).
- The usually accepted first choice test is detection of IgA a-tTG, which displays the highest level of sensitivity (up to 98%) with excellent reproducibility (Porcelli et al., 2016).
- According to Mistler (2015) Anti-deamidated gliadin peptide should be paired with both the total serum IgA and anti-tissue transglutaminase IgA and IgG for screening symptomatic patients on a diet containing gluten for best diagnosis.
- "Patients that test positive by deamidated gliadin peptide and/or anti-tissue transglutaminase should then be confirmed with HLA typing and/or biopsy and started on a gluten-free diet" (Mistler, 2015, p. 214).
- Measurements of antibodies to AGA IgA and IgG, TTG IgA and IgG, ARA IgA, EMA IgA by immunoassay and/or immunofluorescence and testing levels of total IgA include current laboratory choices (Mistler, 2015).
- Genetic testing for HLA-DQ2.5 and HLA-DQ8 haplotypes may be seen as an essential second step in diagnosis (Mistler, 2015).
- The gold standard for diagnosis is biopsy of the small intestine, in which upper gastrointestinal endoscopy is used for obtaining biopsies and should include a minimum of 4 biopsies with two from the distal duodenum and two from the bulbus (Mistler, 2015).

Nursing Care

- Healthcare professionals play an essential role in providing adequate education and support to patients and their families.
- Patients will need to be taught how to identify gluten-free products and how to accurately read food labels.
- Gluten may be used as a stabilizer, emulsifier, and thickening agent in an array of processed foods (O'Donnell, 2016).
- "Many foods are naturally gluten-free, including fresh meats, poultry and fish; eggs; fruits and vegetables; rice; potatoes; milk and cheese; and beans, seeds and nuts in their natural form" (Celiac disease, 2015).
- Patients should also be taught about gluten-free flour options: rice flour, potato starch flour, tapioca starch flour or cornstarch (Celiac disease, 2015).
- Some vitamins and medications may also contain gluten, patients should be encouraged to talk with their local pharmacist to be sure medications do not contain gluten (Celiac disease, 2015).
- Some patients may be at risk for vitamin and mineral deficiencies, including B vitamins, calcium, vitamin D, and iron (Celiac disease, 2015).
- Some patients may need to take a multivitamin or supplement or working closely with a dietician, a proper gluten free diet can be determined to meet nutritional needs (Celiac disease, 2015).
- A multidisciplinary approach carries the highest rate of patient success.
- Nurses need to collaborate with a dietician to teach patients and their families how to eat a balanced diet and adapt to a gluten-free diet with minimal added expense (O'Donnell, 2016).
- The topic of eating out should also be covered, many restaurants now offer gluten-free options and menus (O'Donnell, 2016).
- Cross contamination both at home and when eating out is a topic that should be covered.
- Patients can also be referred to support groups, as patients can support each other in transitioning to and maintaining a gluten-free diet, become educated about the disease, and share information on available resources (O'Donnell, 2016).
- Patients must be encouraged to continue to follow-up with their healthcare providers, as patients with celiac disease are at higher risk for other autoimmune diseases.
- Yearly serologic testing should be completed to ensure that adhering to the gluten-free diet is effective (O'Donnell, 2016).
- Other testing could include bone-density testing, testing for nutritional deficiencies, and also for possible malignancies (O'Donnell, 2016).
- Patients may also be referred to specialists or gastroenterologists for further testing.

Conclusion

Celiac disease is becoming a more prevalent disease and deserves more attention by healthcare professionals. Patients of all ages are affected by the disease and therefore it is essential healthcare professionals examine all signs and symptoms closely. Being familiar with the disease process and possible genetic components is important to proper patient care and diagnosis. Celiac disease is not always easy to diagnose and may be misdiagnosed several times before a proper diagnosis is confirmed. Adequate education and support is essential to patient's lifelong commitment to a gluten free diet and healthy life.

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