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Pathophysiology of Celiac Disease
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Introduction
- My future fiancé-in-law was diagnosed with celiac disease (CD) approximately 2 years ago.
- He was 60 years old at the time of diagnosis and suffered with diarrhea and abdominal pain for a year prior to being told he had CD.
- My fiancé and daughter are at increased risk of developing CD due to the genetic/heritability component of the disease.
- Those who have a first-degree relative with CD have a 4% to 17% increased risk of developing the disease compared to the general population (Cui, Basen, Philipp, Yash, & Krishnanswamy, 2017).

About CD
- CD was first described by Samuel Gee in 1876 and it wasn't until 1941 that celiac was hypothesized as the possible offending agent, noted by William Dike (Leonard, Sepani, Calaboe, & Pimenta, 2011).
- CD was once thought to almost exclusively affect young Caucasian children, but can actually occur at any age and can affect almost anyone (Zindel et al., 2017).
- The prevalence of CD is estimated to be 0.6% to 1% worldwide with approximately 0.7% to 0.8% of the United States (US) population being affected (Cui et al., 2017).
- Non-Caucasians with an increased risk of CD compared to the general population (Cui, et al., 2015).

Underlying Pathophysiology
- CD is an autoimmune disorder occurring in genetically predisposed individuals. Individuals who are genetically predisposed to CD have the HLA-DQ2 or HLA-DQ8 haplotypes (Labovitz et al., 2015).
- While 30-40% of the general population are carriers for at least one of these alleles, the absence of either allele has a “negative predictive value of 100% in excluding the diagnosis of CD” (Green et al., 2015).
- The pathogenesis of CD is dependent not only on being genetically predisposed to the disease, but is also dependent on exposure to gluten and other environmental factors (Green et al., 2015).
- Gluten is the storage protein present in wheat, rye, and barely (Cui et al., 2017).
- It is a mixture of glinid and glutenin, complex proteins that are not completely digested by intestinal enzymes. This triggers an inflammatory process, as these proteins are mistaken as pathogens (Leonard et al., 2017).
- Partially digested glinids then enter through the epithelial barrier of the small intestinal mucosa, triggering innate and adaptive immune responses (Labovitz et al., 2015).
- Once through the epithelial barrier, glinids are “deamidated by the enzyme transglutaminase (TGG)” (Green et al., 2015).
- The process increases the immunogenicity of glinids, by altering the charge that facilitates binding to the HLA-DQ2 or HLA-DQ8 molecules on antigen presenting cells (Labovitz et al., 2015).
- Gladin CD-positive T cells recognize gladin peptides and in turn produce proinflammatory cytokines (Labovitz et al., 2015).
- During the inflammatory cascade, the release of metalloproteinases and other tissue damaging mediators cause tissue injury, leading to epithelial lesions which cause the symptoms seen in the clinical manifestations of CD (Labovitz et al., 2015).
- Other environmental factors have been attributed to the pathogenesis of CD, including infections of the GI tract and alterations in the intestinal microbiota (Green et al., 2015).
- Examples of intestinal alterations include bacteria such as the Bacteroides species, which are thought to play a pathogenic role in the development and manifestation of symptoms in CD (Green et al., 2015).

Significance of Pathophysiology
- The inflammatory process described results in small histologic abnormalities including: “atrophy of the intestinal villi, hyperplasia of the crypts and lymphocytic infiltration of intestinal epithelial spaces and lamina propria” (Adelman et al., 2015).
- The diagnostic test for CD is villous atrophy on biopsy and return of normal villi after adherence to a gluten free diet (Cui et al., 2017).
- Delayed diagnosis of CD showed more severe villous atrophy at diagnosis, which predicted slower muscle healing in the one year follow-up biopsy, as detailed in a study performed by Pelko et al. (2015).
- Undiagnosed CD results in continued ingestion of gluten, which results in continuous inflammation that erodes and flattens the villi, causing insufficient uptake of nutrients as they pass through the intestines (CDF, 2015).

Diagnostic Testing
- In addition to duodenal biopsy, serologic and genetic testing can be performed for diagnosis of CD.
- Serologic testing measures the levels of:
  - Anti-tissue transglutaminase (ITG)
  - Anti-endomysium (EMA)
  - Anti-deamidated gliadin peptides (DGP) (Green et al., 2015).
- First line testing to screen for CD includes measurement of serum IgA antibodies to ITG (Leonard et al., 2017).
- This test has a high sensitivity and specificity and is the first test that should be ordered when CD is suspected. If serum IgA levels are < 0.2, or the patient is younger than 2 years of age, then the test of choice is measurement of IgA levels to DGP (Cui et al., 2017).
- Genetic testing can be considered for patient’s whose serologic and histologic results are inconsistent (Cui et al., 2017).

Implications for Nursing
- CD is increasing in prevalence, although it is generally undiagnosed (Green et al., 2015).
- Therefore, it is important for healthcare providers to recognize not only the GI symptoms present in CD but to also assess the cause of any extra-intestinal findings as well.
  - Patient’s with otherwise unexplained iron-deficiency anemia, as well as those who do not respond to oral iron therapy, should be tested for celiac disease (Leonard et al., 2017).
- CD commonly leads to nutritional deficiencies such as:
  - iron, folic acid, vitamin B12, and B6, copper, zinc and vitamin D deficiencies (Cui et al., 2017).
- Combined diagnoses of CD showed more severe villous atrophy at diagnosis, which predicted slower muscle healing in the one year follow-up biopsy, as detailed in a study performed by Pelko et al. (2015).
- Increased provider awareness about celiac disease could improve healthcare-associated costs by preventing repetitive testing and inadvisability (Robinson et al., 2016).
- Patient education is key in the success of treatment for CD, as strict adherence to a gluten free (GF) diet is the main therapeutic intervention for this disease process (Green et al., 2015).
- Resource should be given to patient’s on how to contact a dietitian upon diagnosis, to low adherence to a GF diet, predisposes patient’s to incomplete muscular recovery, which leads to decreased quality of life due to incomplete resolution of intestinal and extra-intestinal symptoms (Pelko et al., 2015).

Conclusion
- The reason for an increasing prevalence of CD is still unknown, as is the extent to which environmental factors other than gluten contribute to the development of CD (Green et al., 2015).
- The development of CD occurs among subjects with HLA-DQ2 or HLA-DQ8 genes who are exposed to gluten and other environmental factors (Green et al., 2015).
- Diagnosis of CD is based on characteristic autoantibodies seen in serologic testing, as well as with small intestinal histological changes upon endoscopy for patients who have symptoms (2015).
- Early diagnosis can prevent the extent of villous atrophy and extraintestinal complications, as well as the rate of muscular healing and resolution of symptoms seen in patients (Cui et al., 2015).
- Adherence to a strict GF diet is the best predictor of remission of the disease.