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Pathophysiology of Celiac Disease

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Introduction

- My future father-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/hereditary 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, Yusin, & Krishnaswamy, 2017).

About CD

- CD was fist described by Samuel Gee in 1887 and it wasn't until 1941 that wheat was hypothesized as the possible offending agent, noted by William Dicke (Leonard, Sapone, Catassi, & Fasano, 2017).
- CD was once thought to almost exclusively affect young Caucasian children, but can actually occur at any age and can affect almost any race (Leonard 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-Hispanic whites have an approximately 4-8 times greater prevalence of CD compared to other races (Mardini, Westgate, & Grigorian, 2015).

Celiac disease effects **1%** of healthy average Americans. That means at least **3 million** people in our country are living with celiac disease. **97%** of them are undiagnosed.

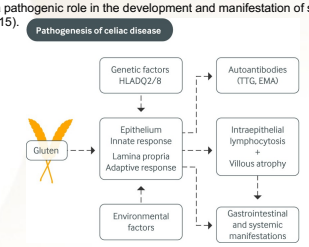
(The University of Chicago Celiac Disease Center, n.d.)

Signs and Symptoms

- The classic presentation of CD manifests as gastrointestinal (GI) symptoms of diarrhea, abdominal pain/distension and malnutrition, resulting in poor growth/failure to thrive in children (Robinson, Davis, Vess, & Lebel, 2015).
- However, the clinical presentation of CD varies from patient to patient with ailments presenting as both intestinal and extra-intestinal symptoms (Green, Lebwohl, & Greywoode, 2015).
- Intestinal symptoms include:
  - diarrhea, bloating, constipation, abdominal pain or weight loss (Leonard et al., 2017).
- Extra-intestinal symptoms include:
  - iron-deficiency anemia, dermatitis herpetifromis, vitamin deficiencies, neurological or psychiatric problems and infertility (Lebwohl, Ludvigsson & Green, 2015).

Underlying Pathophysiology

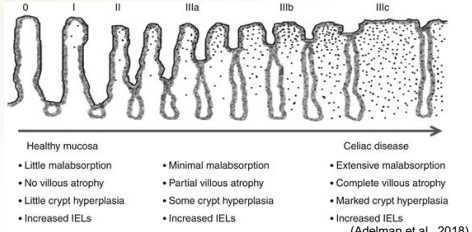
- CD is an autoimmune disorder occurring in genetically predisposed individuals.
- Individuals who are genetically predisposed to CD have the HLA-DQ2 or HLA-DQ8 haplotype (Lebwohl et al., 2015).
- While 30-40% of the general population are carriers for at least one of these alleles, the absence of either 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 gliadin and glutenin's, 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 gliadins then enter through the epithelial barrier of the small intestinal mucosa, triggering innate and adaptive immune responses (Lebwohl et al., 2015).
- Once through the epithelial barrier, gliadins are "deamidated by the enzyme transglutaminase (TTG)" (Green et al., 2015).
- This process increases the immunogenicity of gliadin, by altering the charge that facilitates binding to the HLA-DQ2 or HLA-DQ8 molecule on antigen presenting cells (Lebwohl et al., 2015).
- Gliadin CD4 positive T cells recognize gliadin peptides and in turn produce proinflammatory cytokines (Lebwohl 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 (Lebwohl 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).



(Lebwohl et al., 2015)

Significance of Pathophysiology

- The inflammatory process described results in small histologic abnormalities including:
  - "atrophy of the intestinal villi, hypertrophy of the crypts and lymphocytic infiltration of intraepithelial spaces and lamina propria"(Adelman, et al., 2018).
- Intestinal villi, pictured below, are finger-like projections that line the intestinal walls, allowing absorption and digestion of nutrients. (Celiac Disease Foundation [CDF], 2017).
- Undiagnosed CD, results in the continued ingestion of gluten, which results in continuous inflammation that erodes and flattens the villi, causing inefficient uptake of nutrients as they pass through the intestines (CDF, 2017).
- Diagnosis of CD includes serologic and genetic testing, as well as duodenal biopsy if positive results are found in the initial tests (Cui et al., 2017).
- Duodenal biopsy is performed via upper endoscopy where 2 samples are obtained from the duodenal bulb and at least 4 from the rest of the duodenum (Cui et al., 2017).
- Samples are then examined, mainly assessing the extent of villous atrophy.
- The extent of atrophy is classified based on the Marsh scale, a seven level classification system, with a 0 indicating normal intestinal villi, and a 4 indicating flat, fully atrophied intestinal villi (CDF, 2017).
- The picture below depicts the Marsh Scale Classification in the assessment of villous atrophy.
- The gold standard for diagnosis of CD is villous atrophy on biopsy and return of normal villi after adherence to a gluten free diet (Cui et al., 2017).



(Adelman et al., 2018)

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 (tTG)
  - 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 tTG (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 (Leonard et al., 2017).
- 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 IgG 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 underdiagnosed (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 B_{12} and B_6, copper, zinc and vitamin D deficiencies (Cui et al., 2017).
- Delayed diagnosis of CD showed more severe villous atrophy at diagnosis, which predicted slower mucosal healing in the one year follow-up biopsy, as detailed in a study performed by Pekki et al. (2015).
- Increasing provider awareness about celiac disease could improve healthcare-associated costs by preventing repetitive testing and misdiagnosis (Robinson et al., 2015).
- 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).
- Resources should be given to patient's on how to contact a dietician upon diagnosis, as low adherence to a GF diet, predisposes patient's to incomplete mucosal recovery, which leads to decreased quality of life due to incomplete resolution of intestinal and extra-intestinal symptoms (Pekki 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 examination of biopsy sample (Green et al., 2015).
- Early diagnosis can prevent the extent of villous atrophy seen, which positively correlates to the rate of mucosal healing and resolution of symptoms seen in CD (Pekki et al., 2015).
- Adherence to a strict GF diet is the best predictor of remission of the disease.

References

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