

Celiac Disease: Recognizing and Managing a Multisystem Disorder



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Objectives

Over the last 40 years, the face of celiac disease (CD) has shifted from exclusively intestinal and often malnourished, to include symptoms spanning a range of systems. CD is treatable, but in order to properly screen and diagnose, clinical vigilance is necessary.

- Identify historical factors and clinical findings that indicate screening
- Choose an appropriate diagnostic testing pathway
- Describe clinical management strategies for extraintestinal symptoms

Introduction

- Classic presentation: children with diarrhea, abdominal distension, malabsorption, and failure to thrive
- Emergence of serological testing in 1980s – 90s increased screening and diagnosis
- Now, nonclassical presentations 5-6 x more common
- Extraintestinal presentations = longer time to diagnosis
- 2.3 months v. 3.5 years
- 90% US cases undiagnosed
- Pathophysiology: Autoimmune enteropathy - Ingested gluten disassembles tight junctions, binds to HLA encoded cell surface proteins → T and B lymphocyte activation and producing anti-gliadin and tTG Abs → attack enterocytes, impairing absorption
- Predisposition: Down syndrome, Turner syndrome, Williams syndrome, type 1 diabetes, autoimmune thyroiditis, selective IgA deficiency, 1° relative with CD
- HLA-DQ2 or -DQ8 in ~ 95%
- W>M
- Caucasian > AA, Hispanic



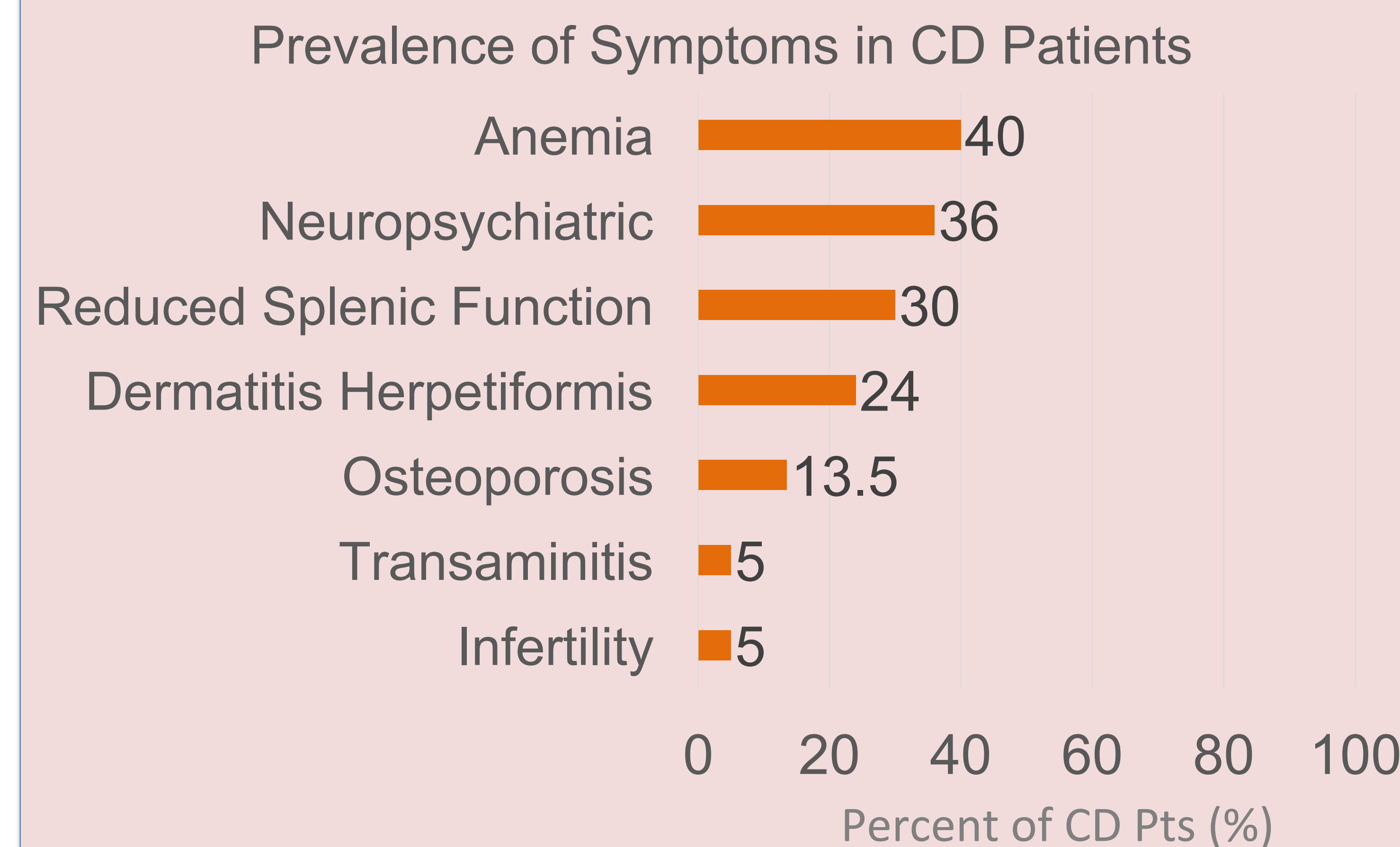
Elson, Boston Children's Hospital Journal, 2005

Intestinal Symptoms

- Mild presentation most common: constipation, diarrhea, dyspepsia and/or nausea.
- PE: Tympanic abdomen, evidence of weight loss
- DDx: IBS, IBD, infectious, SIBO, medication reaction (Sartans)
- **Suspected IBS →screened for CD**
- Overweight/obesity does not r/o CD

Hematologic

- Anemia:** Microcytic, due to malabsorption of iron
- Iron supplementation: May need alternative forms
 - **Refractory IDA →screen for CD**
- Functional hyposplenism:** Due to antibody deposition
- Asymptomatic, anemia, thrombocytosis, fatigue, bleeding, frequent infections
 - Dx: peripheral smear →Howell-Jolly bodies contrast microscopy →pitted RBCs
 - Management: pneumococcal vaccination, annual flu
 - Uncomplicated CD: 1 dose of PCV20 or PCV15 recommended, 1 dose of PPSV23 considered at 1 yr
 - CD + hyposplenism: 1 dose PCV20 or PCV15 and 1 dose of PPSV23



Osteoporosis

- Sx: fractures, kyphosis
- **Idiopathic low BMD →screen for CD**
- Assess on dx and annually: Ca, vit. D, PTH, ALP
- DEXA on dx in elderly/postmenopausal or fracture hx, severe malabsorption, longstanding sx
- Tx: GFD, Ca and vit. D +/- bisphosphonates

OB/GYN

- A/w delayed menarche, early menopause, infertility, intrauterine growth restriction and miscarriage

Transaminitis

- Asymptomatic or fatigue/ malaise, cirrhosis if untreated
- Mild – moderate levels, AST: ALT < 1; Tx: GFD, monitor
- **Autoimmune hepatitis →screen for CD**

Dermatitis Herpetiformis

- Symmetrical, polymorphic lesions; erythema, urticarial plaques, and papules
- Extensor surfaces, shoulders, buttocks, sacral region, neck, face, scalp
- Untreated →bullous pemphigoid
- Often sole symptom, DDx: eczema
- Tx: GFD, dapsone, steroids, sulfones



Peraza, Merck Manual 2022

Neuropsychiatric

- Peripheral neuropathy:** Numbness, paresthesias, pain; distal → proximal; DDx: DM neuropathy
- Ataxia:** Poor control of extremities, poor coordination, gait instability, speech and vision dysfunction; Tx: IVIG if refractory to GFD
- Epilepsy:** Complex partial sz most common, occipital or temporal focus; Tx= GFD, folate, antiepileptics
- Psych:** A/w anxiety, depression, panic disorder, schizophrenia

Diagnostic Tools



CME Questions

1. What is the most common cutaneous manifestation of CD?
 - a. Eczema
 - b. Dermatitis herpetiformis
 - c. Psoriasis
 - d. Erythema nodosum
2. CD impacts the vaccination schedule of which vaccine(s)?
 - a. Rotavirus
 - b. Flu and COVID
 - c. Meningococcal and Pneumococcal
 - d. Shingles
3. Which presentation should prompt CD screening?
 - a. Anemia unresponsive to oral supplementation
 - b. Osteopenia in a woman over 65 years old
 - c. Elevated hemoglobin and abdominal pain
 - d. Peripheral neuropathy in the setting of diabetes

Answers: B, C, A

References:

1. Caio G, Volta U, Sapone A, et al. Celiac disease: a comprehensive current review. BMC Medicine. 2019;07/23 2019;17(1):142. doi:10.1186/s12916-019-1380-z
2. Laurikka P, Nurminen S, Kivela L, Kurppa K. Extraintestinal Manifestations of Celiac Disease: Early Detection for Better Long-Term Outcomes. Nutrients. Aug 3 2018;10(8):doi:10.3390/nu10081015
3. Lewohl B, Rubio-Tapia A. Epidemiology, presentation, and diagnosis of celiac disease. Gastroenterology. 2021;160(1):63-75.
4. Rubio-Tapia A, Hill ID, Semrad C, et al. American College of Gastroenterology Guidelines Update: Diagnosis and Management of Celiac Disease. Am J Gastroenterol. Jan 1 2023;118(1):59-76. doi:10.14309/ajg.0000000000002075
5. Seidita A, Mansueti P, Compagnoni S, et al. Anemia in Celiac Disease: Prevalence, Associated Clinical and Laboratory Features, and Persistence after Gluten-Free Diet. Journal of Personalized Medicine. 2022;12(10):1582.
6. Salmi T, Hervonen K, Reunala T. Chapter 8 - Dermatitis herpetiformis – a cutaneous manifestation of celiac disease. In: Schieptli A, Sanders D, eds. Celiac Disease and Gluten-Related Disorders. Academic Press; 2022:161-177.
7. Lungaro L, Manza F, Costanzini A, et al. Osteoporosis in Celiac Disease: An Update. Preprints.org; 2022.
8. Villavicencio Kim J, Wu GY. Celiac Disease and Elevated Liver Enzymes: A Review. J Clin Transl Hepatol. Feb 28 2021;9(1):116-124. doi:10.14218/JCTH.2020.00089
9. Balaban DV, Popp A, Ionita Radu F, Jinga M. Hematologic manifestations in celiac disease—a practical review. Medicina. 2019;55(7):373.
10. Trovato CM, Raucci U, Vallutti F, et al. Neuropsychiatric manifestations in celiac disease. Epilepsy & Behavior. 2019;100/1/ 2019;99:106393. doi:https://doi.org/10.1016/j.yebeh.2019.06.036