

Celiac Disease	
<b>PowerPlans</b>	Celiac Disease Orderset, Celiac Gene Assessment, GI AMB Celiac Disease (Future) Plan
<b>Presentation</b>	<ul style="list-style-type: none"> <li>• <b>Classical:</b> Malabsorption (FTT, steatorrhea), abd pain, gas, <b>distension, constipation or diarrhea</b>, anemia, non-erosive arthritis, dental enamel defects, aphthous ulcers, dermatitis herpetiformis (pruritis papules/vesicles), neuropsych (ADHD, depression, HA). ↑ risk in T1DM, autoimmune thyroid dz, Turner and Down syndrome.</li> <li>• <b>Infants:</b> <b>present irritable, wasted extremities, buttocks, and distended abdomen</b></li> </ul>
<b>Pathophys</b>	<b>HLA-DQ2 or -DQ8</b> (predisposition, <b>necessary for dz</b> ) + environmental trigger → <b>Antibodies to gliadin</b> (gluten byproduct), tissue transglutaminase (tTG; cross-links and deamidizes gliadin peptides) → <b>enterocyte destruction</b>
<b>Workup</b>	<ul style="list-style-type: none"> <li>• <b>Serologies:</b> anti-tTG IgA, anti-endomysial IgA, anti-gliadin. Always check IgA levels (IgA deficiency can yield false-negatives); DGP IgA if &lt; 2 yrs old</li> <li>• <b>Biopsy:</b> intraepithelial lymphocytes, villous atrophy, crypt hyperplasia</li> </ul>
<b>Treatment</b>	<ul style="list-style-type: none"> <li>• Gluten-free diet (\$\$, needs very strict adherence, hard to maintain). Wheat, rye, barley all contain gluten. Oats are controversial. Improvement in 2-4w.</li> <li>• <b>Follow with TTG until normalized (usually by 12 months). Follow Vitamin D and B 12 levels as well as Thyroid.</b> Check if immune against Hep B</li> </ul>

Malabsorption		
	Presentation/Pathophys	Workup
<b>Carbs</b>	<ul style="list-style-type: none"> <li>• <b>Frequent, watery stools</b></li> <li>• Pathophys: carbs digested by amylase (saliva and pancreas), so <b>pancreatic disease</b> can lead to poor carb digestion</li> <li>• <b>Lactase deficiency</b> (lactose intolerance): usually adult-onset</li> <li>• <b>Bacterial overgrowth</b>/alteration of bowel flora → increased lactate production and temporary lactase deficiency leading to lactose intolerance</li> </ul>	<ul style="list-style-type: none"> <li>• Fecal pH &lt; 5.5 (can also be seen transiently in viral enteritis)</li> <li>• Stool reducing substances &gt;0.5%. *need fresh stool</li> <li>• Breath hydrogen test used to detect lactase and sucrase deficiency (rare)</li> </ul>
<b>Fat</b>	<ul style="list-style-type: none"> <li>• <b>Greasy, foul-smelling stools</b> (steatorrhea)</li> <li>• Cause by diseases affecting bile production/secretion or <b>poor enterohepatic circulation of bile salts</b> (e.g., ileal resection) or <b>pancreatic insufficiency</b> (eg. cystic fibrosis, Schwachman-Diamond) 2/2 inadequate lipase</li> <li>• Critically affects absorption of the fat-soluble vitamins <b>A, D, E, and K</b>.</li> <li>• <b>Giardia</b> infection often associated with fat malabsorption</li> </ul>	<ul style="list-style-type: none"> <li>• Spot fecal fat: non-specific</li> <li>• Split fats (fatty acids) more suggestive of malabsorptive process</li> <li>• Neutral fats more suggestive of pancreatic dysfunction</li> <li>• 72 hr fecal fat: &gt; 5 g per 24 hours suggests malabsorption (diet during these 24 hrs should be &gt;35% fat)</li> </ul>
<b>Protein</b>	<ul style="list-style-type: none"> <li>• <b>Edema, hypoalbuminemia</b></li> <li>• Usually related to <b>deficiency of pancreatic proteases (e.g. cystic fibrosis)</b></li> <li>• Different from protein-losing gastroenteropathy (PLE) (2/2 mucosal disruption or increased lymphatic pressure)</li> </ul>	<ul style="list-style-type: none"> <li>• Serum total protein, albumin</li> <li>• Stool alpha-1 antitrypsin (for PLE)</li> </ul>

Autoimmune Hepatitis	
<b>Presentation</b>	Acute vs. subacute. Transaminitis > bilirubin elevation. Hypergammaglobulinemia. Fatigue, amenorrhea.
<b>Pathophys</b>	<ul style="list-style-type: none"> <li>• <b>Type 1 (classic):</b> any age/gender. +ANA, anti-SM.</li> <li>• <b>Type 2:</b> girls. anti-LKM. Recurrence more common in Type 2.</li> </ul>
<b>Workup</b>	LFTs, Ig levels, auto-antibodies, maybe liver biopsy
<b>Treatment</b>	Prednisone (18-24m) + azathioprine/6-MP (steroid-sparing; check TPMT enzyme activity first. Low TPMT levels = risk of myelosuppression). Relapse more common if tx weaned in 1st 3 years of therapy or during puberty.