

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

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

Autoimmune Hepatitis	
Presentation	Acute vs. subacute. Transaminitis > bilirubin elevation. Hypergammaglobulinemia. Fatigue, amenorrhea.
Pathophys	<ul style="list-style-type: none"> • Type 1 (classic): any age/gender. +ANA, anti-SM. • Type 2: girls. anti-LKM. Recurrence more common in Type 2.
Workup	LFTs, Ig levels, auto-antibodies, maybe liver biopsy
Treatment	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.