



# Small and Large Intestinal pathology, part 1

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# Diseases of the intestines



- ▶ Intestinal obstruction
- ▶ Vascular disorders
- ▶ Malabsorptive diseases and infections
- ▶ Inflammatory bowel disease.
- ▶ Polyps and neoplastic diseases

# Intestinal obstruction

- ▶ **Mechanical obstruction:**

- ▶ Intussusception

- ▶ Hernias.

- ▶ Adhesions.

- ▶ Volvulus

- ▶ Tumors.

- ▶ Diverticulitis

- ▶ Infarction

- ▶ **Non-mechanical obstruction**

- ▶ Hirschsprung disease

- ▶ Neurological disorders.

- ▶ Drugs....etc



# Clinical picture of intestinal obstruction.

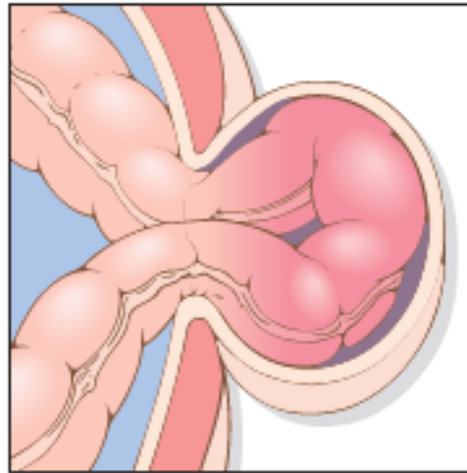
- ▶ Abdominal pain
- ▶ Distention
- ▶ Vomiting
- ▶ Constipation.
  
- ▶ Acute or chronic.



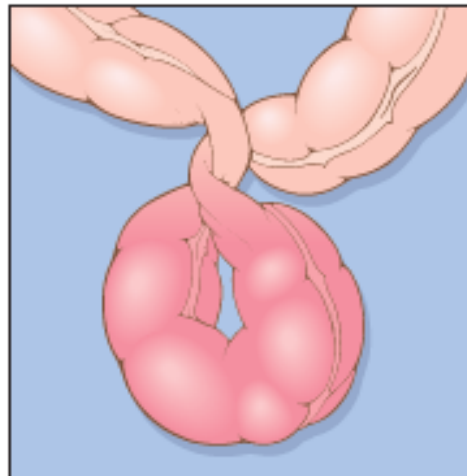
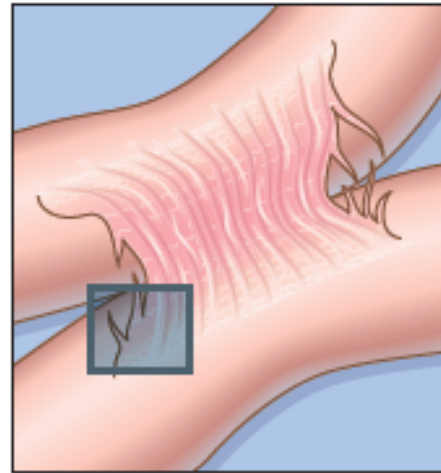
# 80% of mechanical obstructions



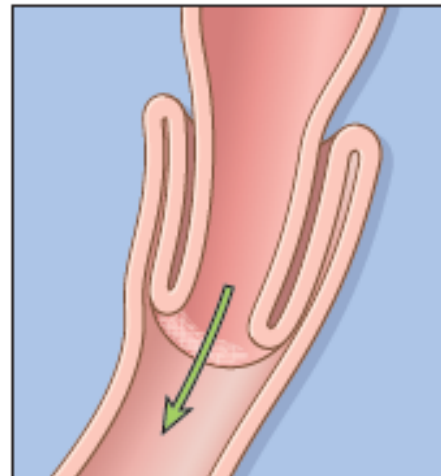
**Herniation**



**Adhesions**



**Volvulus**



**Intussusception**



# Intussusception

- ▶ Segment of the intestine constricted by a wave of peristalsis, telescopes into the immediately distal segment.
- ▶ Once trapped, invaginated segment is propelled by peristalsis, and pulls mesentery with it.
- ▶ **Most common cause of intestinal obstruction in children younger than 2 years of age.**
- ▶ Untreated progresses to infarction.

# Causes of intussusception



- ▶ < 2years : **Idiopathic in most cases.**
- ▶ Peyer patches hyperplasia (rotavirus vaccine, viral infections)
- ▶ Meckles diverticulum (ileum)
- ▶ Old children & adults: Intraluminal mass or tumors

# Clinical features:



- ▶ **Abdominal swelling**
- ▶ **Vomiting**
- ▶ **Passing stools mixed with blood and mucus (currant jelly stool)**
- ▶ **Pain.**



# Management

- ▶ Contrast enemas in uncomplicated idiopathic cases.
- ▶ Surgery if complicated or if masses are the leading point.



# Hirschsprung Disease

- ▶ Congenital defect in colonic innervations
- ▶ Congenital aganglionic megacolon
- ▶ More common in males
- ▶ More severe in females
- ▶ Risk increase in siblings.
  
- ▶ **Typical presentation:**
  
- ▶ Neonatal failure to pass meconium
- ▶ Obstructive constipation.



# Pathogenesis



- ▶ **During embryogenesis**
- ▶ Disrupted migration of neural crest cells from cecum to rectum.
- ▶ **Lack of Meissner submucosal plexus and the Auerbach myenteric plexus.**
- ▶ Failure of coordinated peristaltic contractions.
- ▶ Mutations in RET: in familial cases and 15% of sporadic
- ▶ Other genes and environmental factors play role.

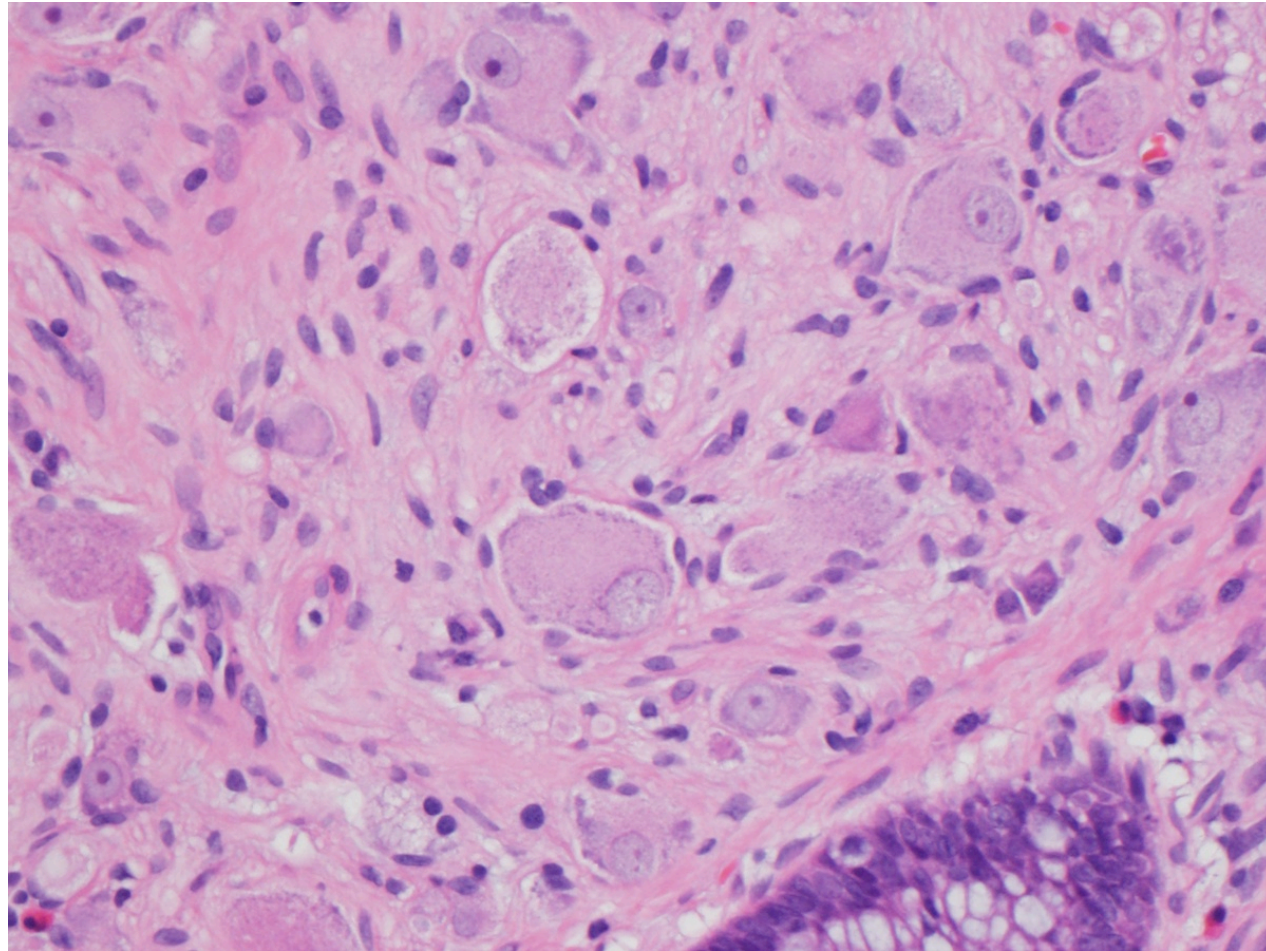
# Morphology



- ▶ Rectum always involved.
- ▶ Extent is variable.
- ▶ Most cases in rectosigmoid.
  
- ▶ **Macroscopic**
- ▶ Aganglionic region normal or contracted
- ▶ Proximal normal segment progressively dilated.
  
- ▶ **Diagnostic workup: barium enema, BIOPSY, microscopic.**



# ganglion cells



<http://www.pathologyoutlines.com>





# Complications

- ▶ Enterocolitis
- ▶ Fluid and electrolyte disturbances
- ▶ Perforation
- ▶ Peritonitis
  
- ▶ **Treatment:**
- ▶ Surgical resection of aganglionic segment and anastomosis of normal segments.

# VASCULAR DISORDERS OF BOWEL



- ▶ **Ischemic Bowel Disease**
- ▶ **Hemorrhoids**





# Hemorrhoids

- ▶ Dilated anal and perianal collateral vessels that connect the portal and caval venous systems.
- ▶ **Predisposing factors:**
  - ▶ Constipation and straining
  - ▶ Venous stasis of pregnancy,
  - ▶ Portal hypertension.
- ▶ External and internal hemorrhoids



- ▶ Thin -walled, dilated, submucosal vessels beneath anal or rectal mucosa.
- ▶ **Symptoms:**
- ▶ Bleeding, pain, thrombosis and inflammation

# DIARRHEAL DISEASE

- ▶ Diarrhea: increase in stool mass, frequency or fluidity.
- ▶ Dysentery: painful , bloody, small volume diarrhea.
  
- ▶ **Malabsorptive Diarrhea**
- ▶ Pancreatic insufficiency.
- ▶ Celiac disease
- ▶ Crohn disease
- ▶ Cystic Fibrosis
- ▶ Lactase (Disaccharidase) Deficiency
- ▶ Abetalipoproteinemia
  
- ▶ **Infectious Enterocolitis**
- ▶ **Inflammatory bowel diseases.....**



# Malabsorptive Diarrhea



- ▶ **Chronic.**
- ▶ Defective absorption of fats, fat- and water-soluble vitamins, proteins, carbohydrates, electrolytes, minerals and water
- ▶ **Hallmark is : steatorrhea.**



# Malabsorptive diarrhea

## Defect in one of the following:

- ▶ Intraluminal digestion.
- ▶ Terminal digestion.
- ▶ Transepithelial transport.
- ▶ Lymphatic transport.

# Manifestations:



- ▶ Weight loss, anorexia,
- ▶ Flatus, abdominal distention,
- ▶ Borborygmi, Muscle wasting
- ▶ Anemia and mucositis (iron, pyridoxine (VB6), folate, or vitamin B12 deficiency)
- ▶ Bleeding (vitamin K deficiency)
- ▶ Osteopenia and tetany (calcium, magnesium, or vitamin D deficiency)
- ▶ Neuropathy (vitamin A or B12 deficiency)
- ▶ Skin and endocrine disorders.

# Cystic Fibrosis



- ▶ Mutations in cystic fibrosis transmembrane conductance regulator (CFTR)
- ▶ Defects in ion transport across intestinal and pancreatic epithelium.
- ▶ Thick viscous secretions.
- ▶ Mucus plugs in pancreatic ducts >>> pancreatic insufficiency (80% of patients).
- ▶ Defect in intraluminal digestion.

# Celiac Disease

- ▶ *Gluten sensitive enteropathy*
- ▶ Immune mediated enteropathy
- ▶ Wheat, rye or barley.
- ▶ Genetically predisposition, HLA-DQ2 or HLA-DQ8.
- ▶ Treatment: gluten free diet.
  
- ▶ Association with: type 1 diabetes, thyroiditis, and Sjogren syndrome

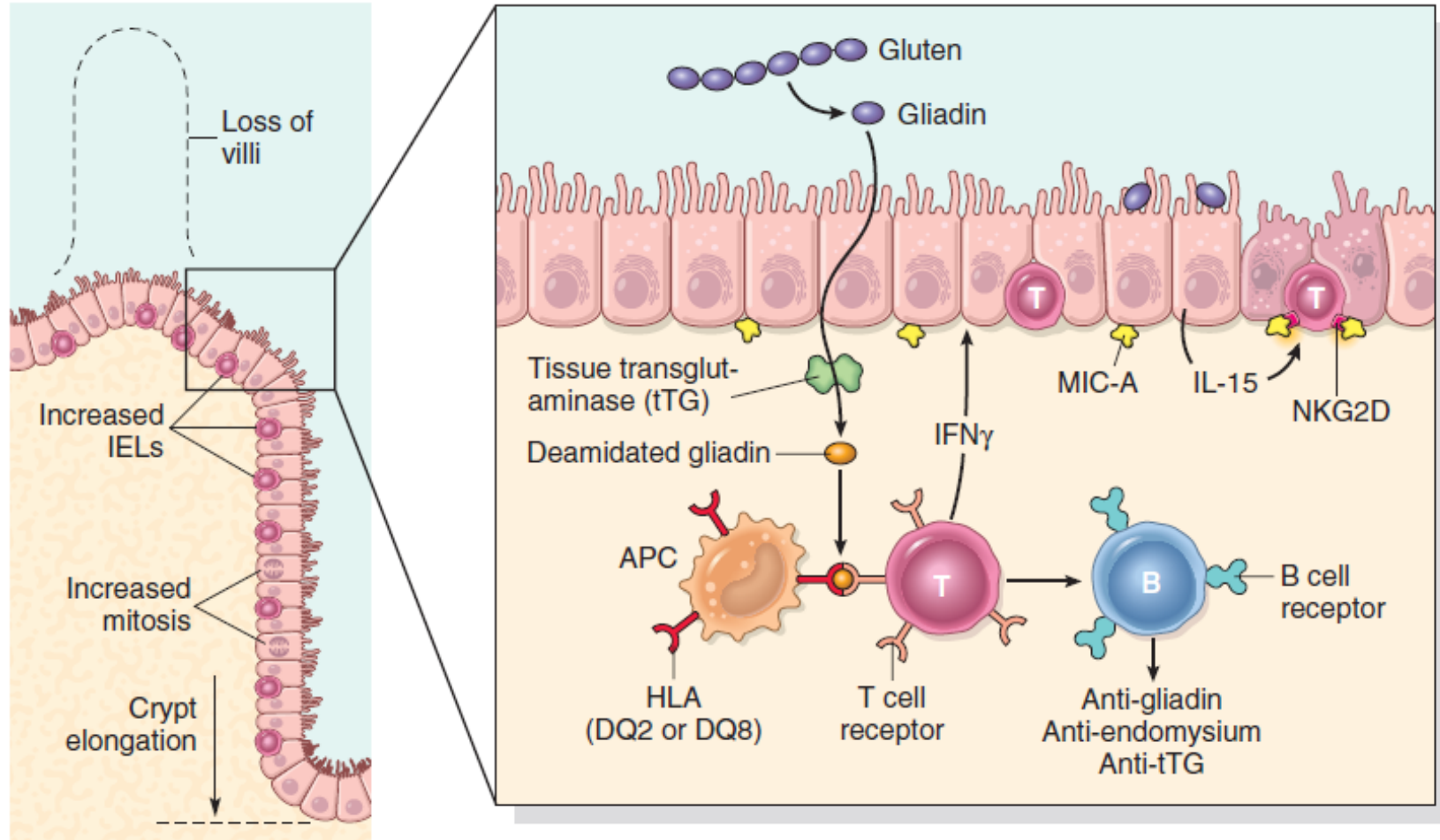




# Pathogenesis

- ▶ Gluten >>> gliadin >>> react with HLA-DQ2 or HLA-DQ8 on antigen-presenting cells >>> CD4+ T cells activation >>> cytokines >>> tissue damage.
- ▶ Serology:
  - ▶ Anti- tissue transglutaminase antibodies
  - ▶ Anti-gliadin antibodies.
  - ▶ Anti -endomysial antibodies



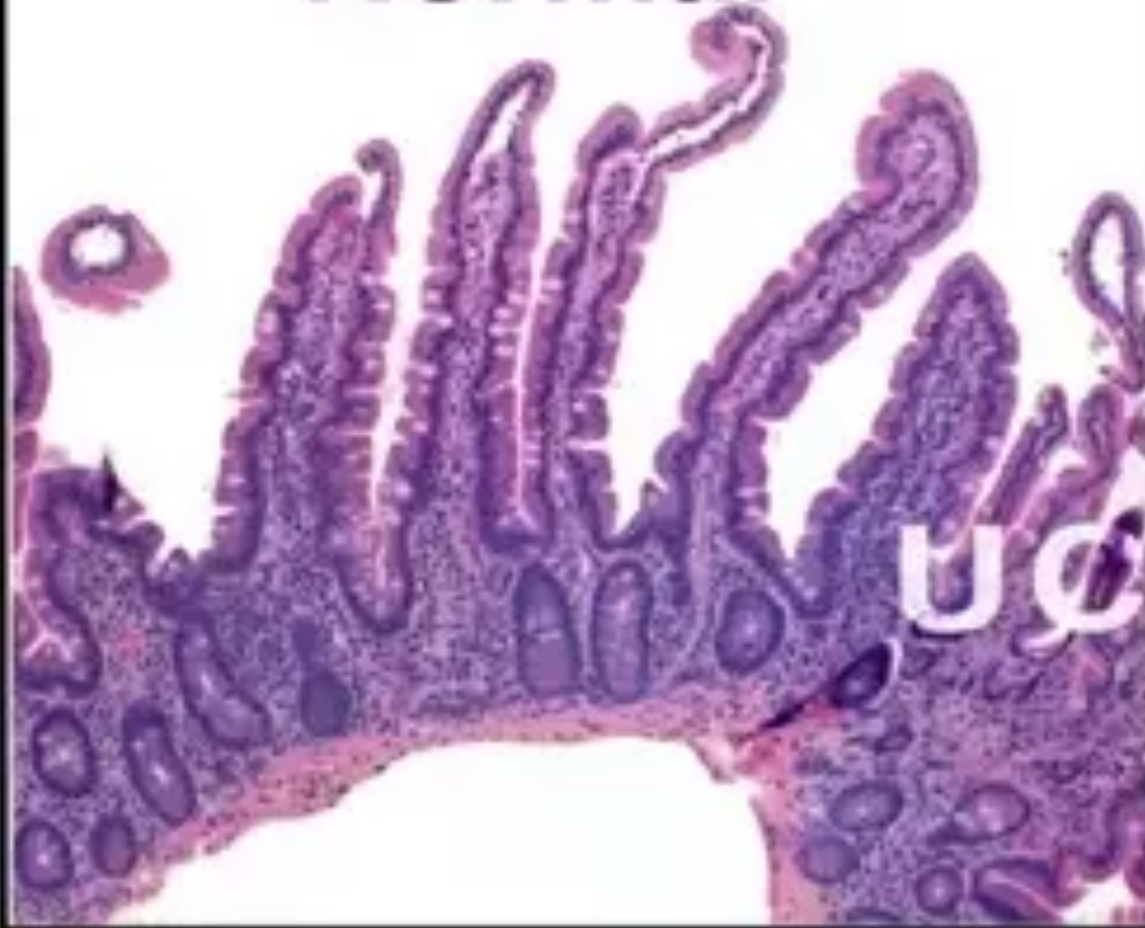


# MORPHOLOGY

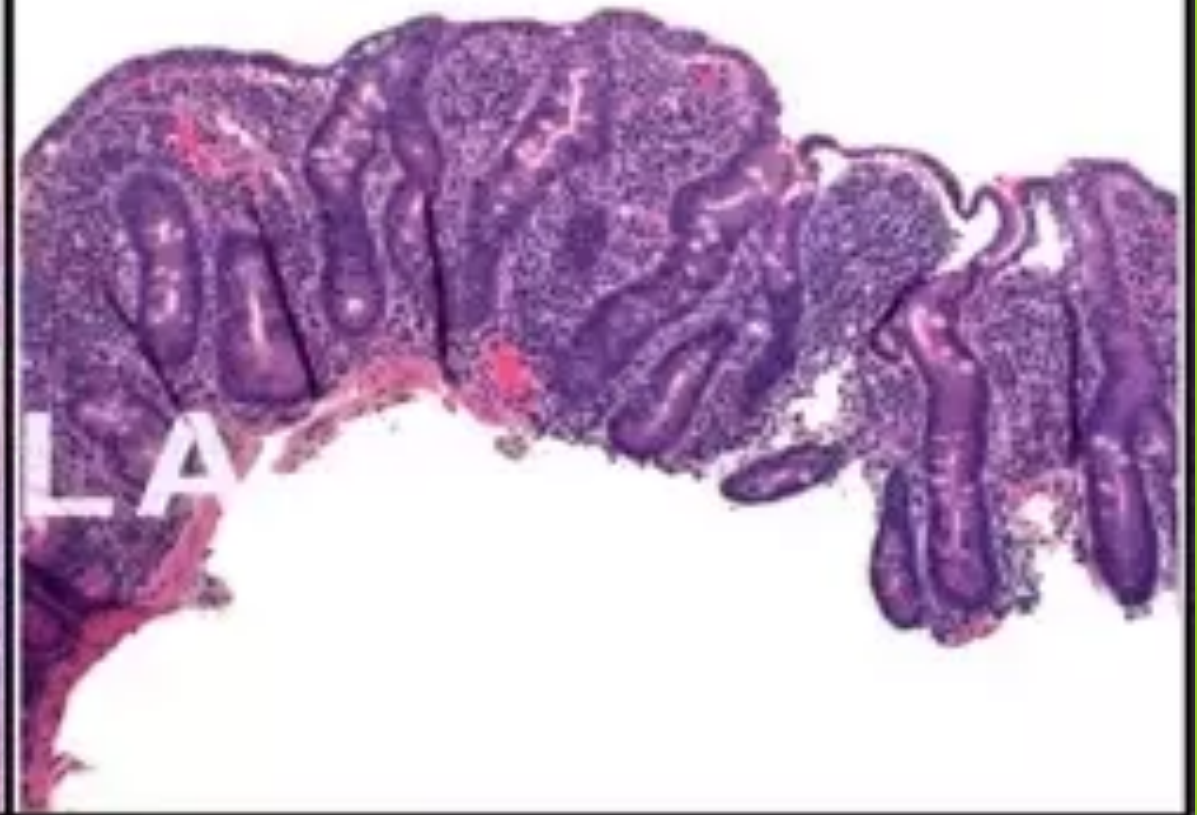
- ▶ Second portion of the duodenum or proximal jejunum.
- ▶ **Triad:** intraepithelial lymphocytosis (CD8+ T cells), crypt hyperplasia, and villous atrophy.
- ▶ Lamina propria: lymphocytes, plasma cells, eosinophils.....
- ▶ IEL & villous atrophy are not pathognomonic, seen in viral enteritis.
  
- ▶ **Diagnosis:** Clinical, histologic and serologic correlation.



**Normal**



**Celiac Disease**



UCLA



# Clinical Features

- ▶ **Children 6-24 months : classical or non classical symptoms**
- ▶ **Classical:** Irritability, abdominal distention, anorexia, diarrhea, failure to thrive, weight loss, or muscle wasting
- ▶ **Non-classical:** abdominal pain, nausea, vomiting, bloating, or constipation.
- ▶ Blistering skin lesion, **dermatitis herpetiformis**, in 10% of Pnts.



# Dermatitis herpetiformis.





- ▶ Adults (30-60 years)
  - ▶ Anemia: iron deficiency
  - ▶ B12 and folate deficiency: less common.
  - ▶ Diarrhea , bloating, and fatigue.
  - ▶ Missed diagnosis: Silent celiac or latent celiac.
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- ▶ Increased risk of enteropathy associated T cell lymphoma & Small intestinal adenocarcinoma

# Diagnosis:

- ▶ **Non invasive serologic tests:**
- ▶ **Most sensitive:**
- ▶ Anti tissue transglutaminase antibody, IgA
- ▶ Anti deamidated gliadin antibodies, IgA & IgG
  
- ▶ **Most specific, but less sensitive**
- ▶ Antiendomysial antibody.
  
- ▶ **Invasive tests: small bowel biopsy.**





# Lactase (Disaccharidase) Deficiency

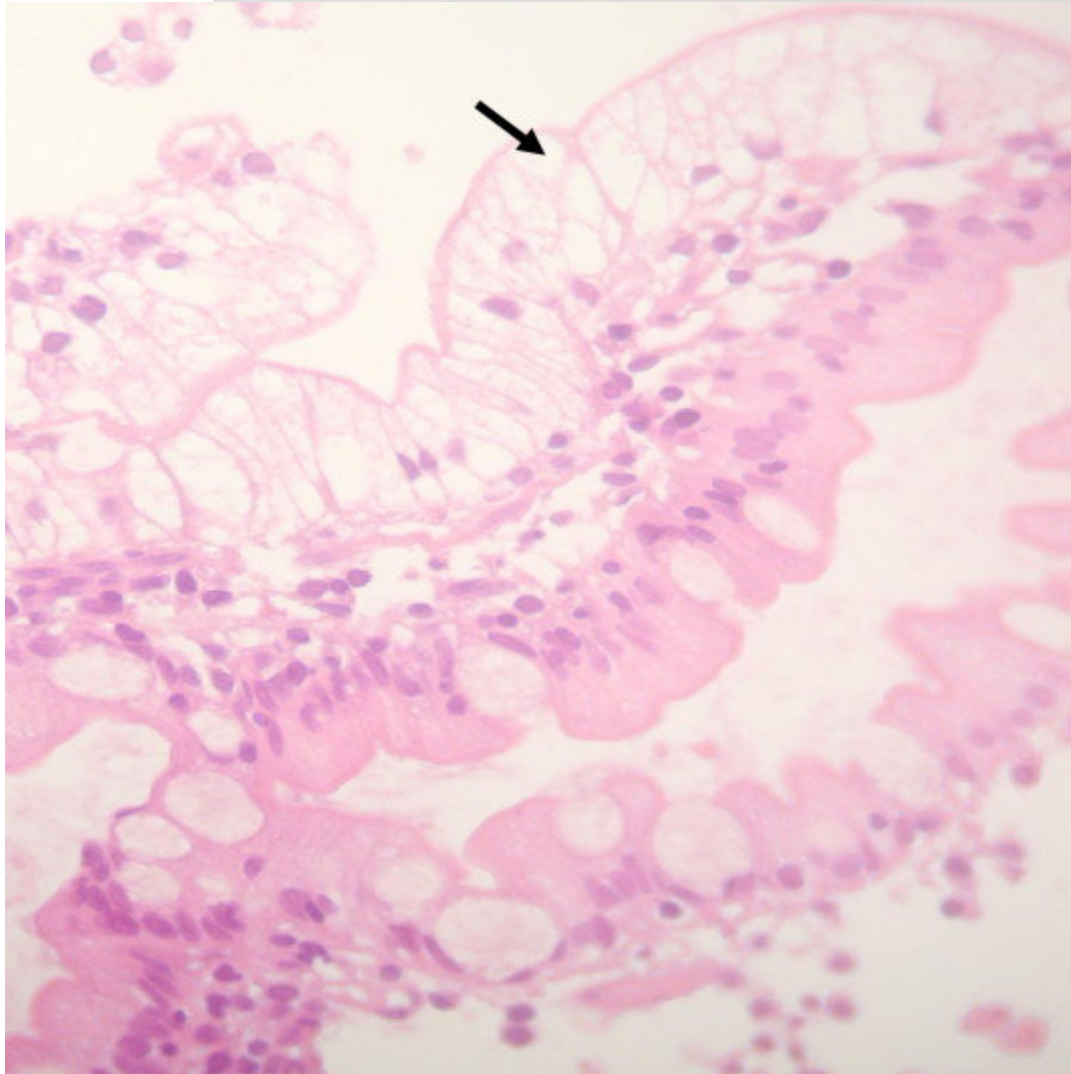
- ▶ Osmotic diarrhea
- ▶ Lactose remains in the gut lumen.
- ▶ Lactase found at apical brush border membrane
- ▶ Normal biopsy findings.
- ▶ Two types:
  - ▶ ***Congenital*** : AR, genetic mutation, rare, explosive diarrhea, watery, frothy stools & abdominal distention, after milk ingestion
  - ▶ ***Acquired*** : follow viral or bacterial enteritis, downregulation of gene, after childhood.



# Abetalipoproteinemia

- ▶ Autosomal recessive, rare.
- ▶ Infants w/ failure to thrive, diarrhea, and steatorrhea
- ▶ Lack of absorption of fat and fat soluble vitamins
  
- ▶ Inability to synthesize triglyceride-rich lipoproteins.
- ▶ Transepithelial transport defect of TG and FAs.
- ▶ Monoglycerides and triglycerides accumulate in epithelial cells.





Micrograph showing enterocytes with a clear cytoplasm (due to lipid accumulation) characteristic of abetalipoproteinemia.

