# Clinical presentation and diagnosis of inflammatory bowel disease in children

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### Introduction

- inflammatory bowel disease (IBD) is comprised of two major disorders: ulcerative colitis (UC) and Crohn disease (CD). UC affects the colon.
- CD can involve any component of the gastrointestinal tract from the oral cavity to the anus.
- Peak incidence in adolescents and young adults.

# Diagnostic evaluation of IBD involves five steps

- Clinical suspicion of the illness
- Exclusion of other illnesses
- Differentiation between CD and UC
- Localization of the region of the disease
- Identification of extraintestinal manifestations

Patients with very early-onset IBD (ie, under age six years, and especially those under age two years) may also warrant immunologic evaluation and genetic sequencing for monogenic forms of IBD.

### **Epidemiology**

- The peak incidence of IBD occurs in patients between the ages of 15 and 30 years [1]. Approximately 5 to 10 percent of patients develop their IBD during childhood or adolescence
- Compared with adults, children with IBD are more likely to present with extensive intestinal involvement and have rapid clinical progression
- family history of IBD, suggesting a stronger genetic association

### **CLINICAL MANIFESTATIONS**

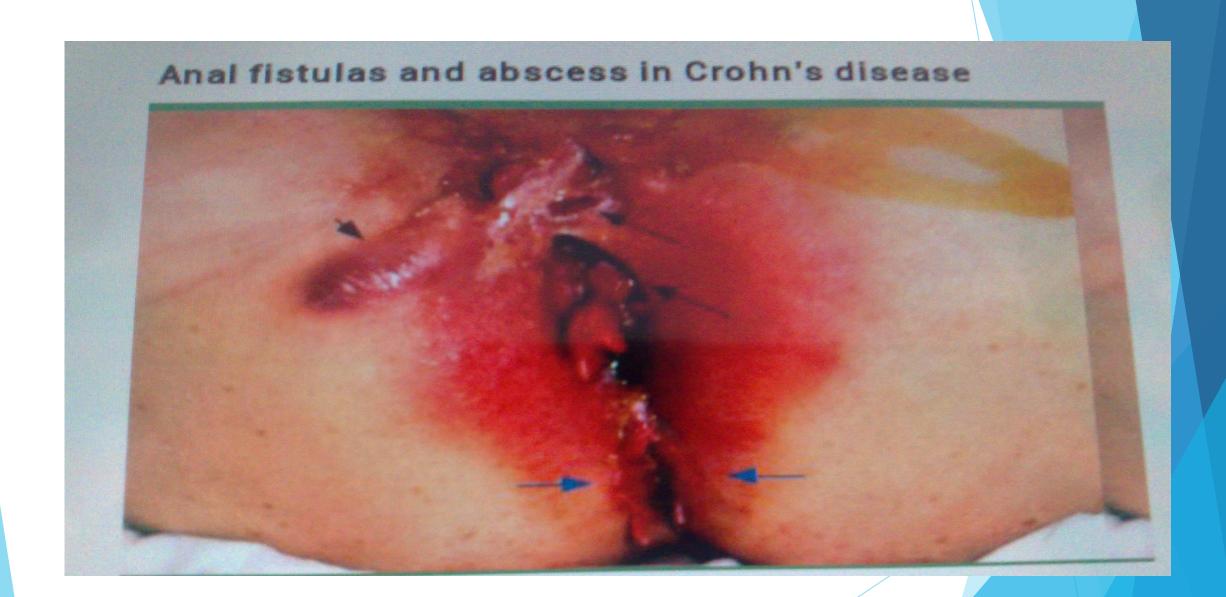
- Gastrointestinal symptoms
- Growth Growth failure
- Physical findings Abdominal tenderness or mass perianal disease (fistulae, anal skin tags, or fissures, or occult blood in stool.
- Systemic symptoms Fever and fatigue
- Extraintestinal manifestations Oral ulcerations, clubbing, rash (erythema nodosum or pyoderma gangrenosum), eye inflammation (uveitis), jaundice or hepatomegaly, or arthritis

# Other gastrointestinal manifestations

- Nonspecific gastrointestinal symptoms
- abdominal pain. The most useful indicators of possible IBD are diarrhea, growth failure, pubertal delay, weight loss, rectal bleeding, pallor/fatigue, perianal skin tags, perianal fistulae or abscesses, a palpable abdominal mass, and a family history of IBD. The abdominal pain associated with CD is often focal and located in the right lower quadrant.

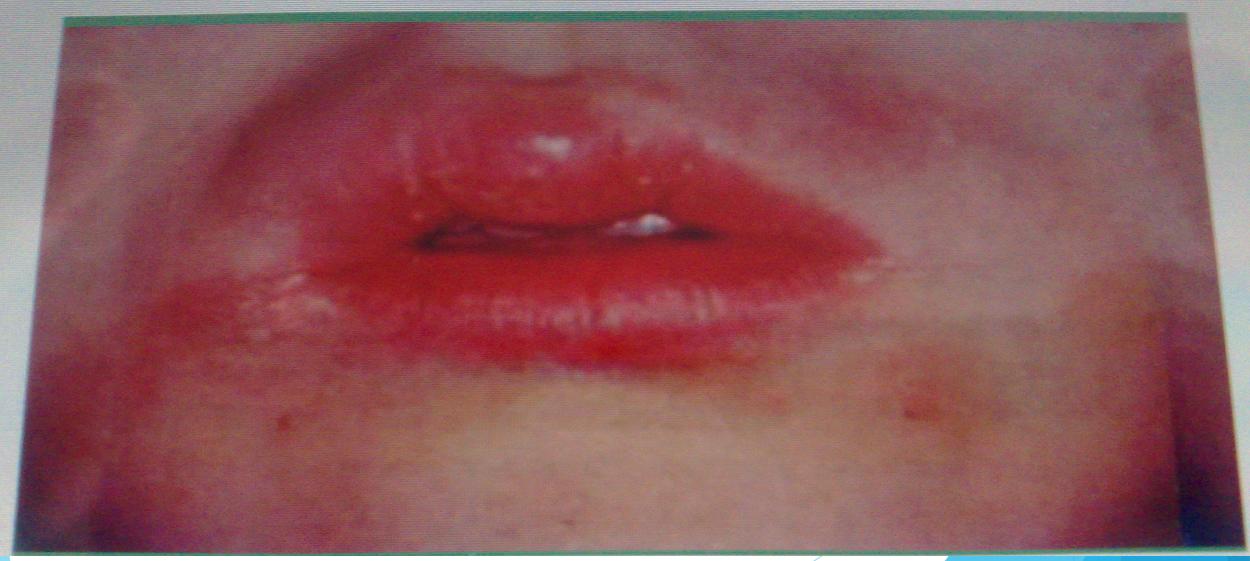
### **Extraintestinal manifestations**

- ▶ 10 percent of patients at presentation and up to 30 percent of patients within the first few years after the IBD is diagnosed . the mouth, skin, joints, liver, eye, and (rarely) other organs
- Idiosyncratic more often associated with colonic disease
- more common in CD oral aphthae (superficial ulcerations, also known as aphthous stomatitis), erythema nodosum, digital clubbing

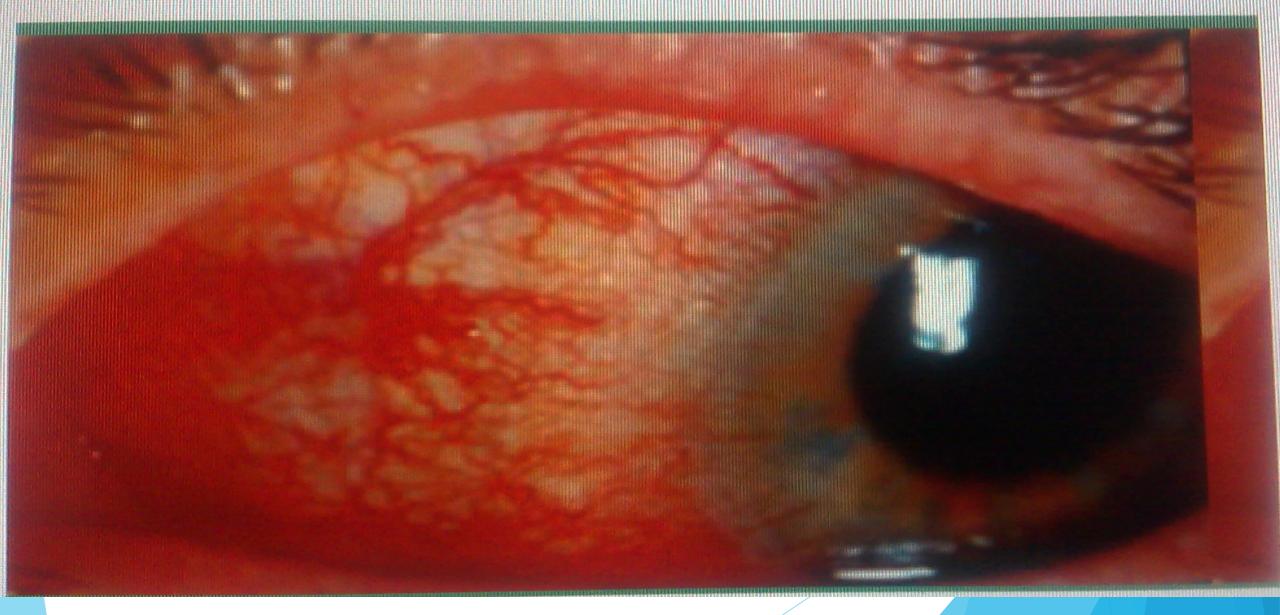




### Lip swelling with fissures



### Diffuse episeleritis



# Anterior uveitis in a patient with inflammatory disease





### <u>Lyodennagairg felicialin</u>





### **Laboratory features**

- Blood tests
- Anemia, increased white blood cell, and platelet count 70%, hemoglobin level <11 g/L</p>
- Elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) 65 to 75 percent
- CRP was elevated in 100 percent of those with CD and 60 percent of those with UC
- Depressed albumin level 40 percent (eg, <3.4 g/L)</p>

Normal blood laboratory tosts do not evolude the diagnosis of IRD	
Normal blood laboratory tests do not exclude the diagnosis of IBD.	

### **Stool tests**

- Gross or occult blood
- Fecal calprotectin
- High sensitivity
- Low specificity (false- positive rate of 30 percent) (eg, >200 mc/g)
- Elevated levels of fecal calprotectin are also found in other causes of inflammatory diarrhea including bacterial and viral enteritis, intestinal lymphoma, celiac disease, food allergy and immunodeficiency. Juvenile Polyps.

## Fecal calprotectin levels are correlated with disease activity in both CD and UC

- Fecal lactoferrin
- Fecal leukocytes

# CANDIDATES FOR EVALUATION Clinical suspicion

- Bloody diarrhea
- Growth failure
- Chronic watery diarrhea
- Chronic abdominal pain
- Perianal abscesses, fistulae, and fissures, oral ulcers, or arthritis
- Laboratory abnormalities

### **DIAGNOSTIC EVALUATION**

- Stool testing (for enteric pathogens below)
- Small bowel imaging
- Upper endoscopy and colonoscopy

### **Imaging**

- MRE is the preferred small bowel imaging modality in children, The main advantage is avoidance of radiation exposure . sensitivity of 83 percent and a specificity of 93 percent
- intraabdominal or perianal fistula or abscess
- Liver , kidney bone disease
- MRE combines oral contrast distension of the bowel with intravenous administration of a gadolinium-based contrast material

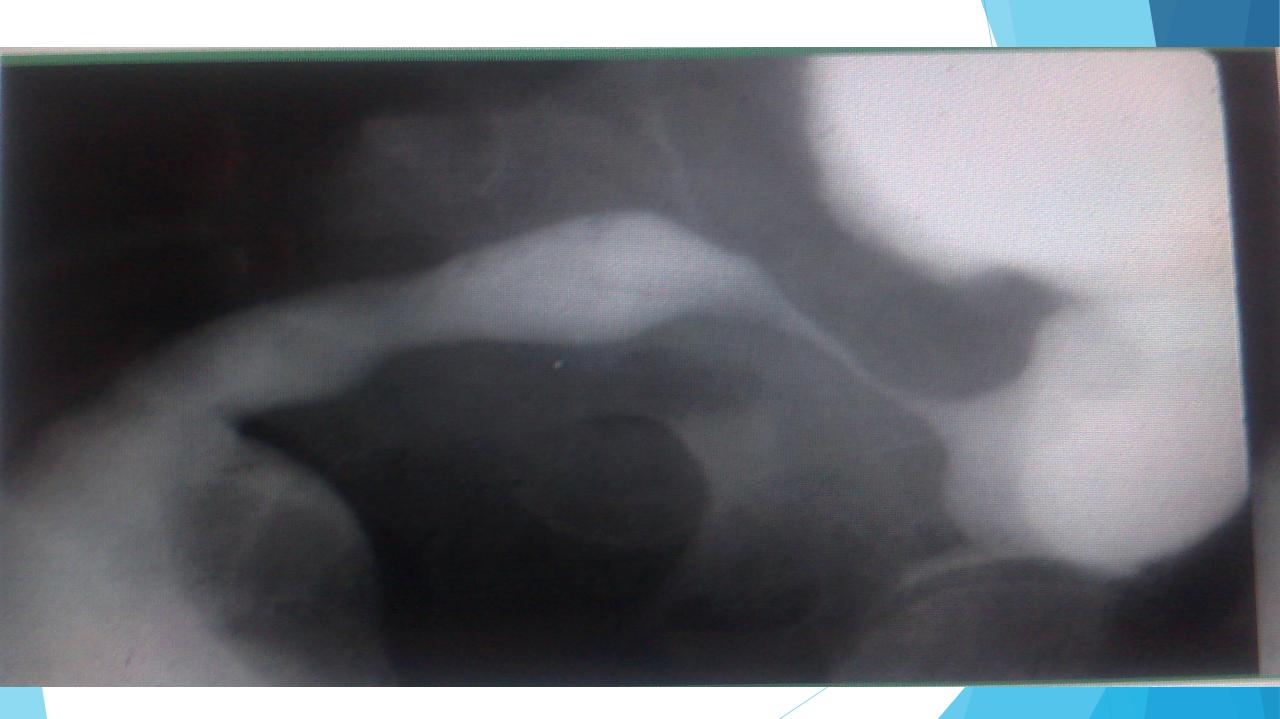
### OT scan of ulcerative colitis





### Chronic ulcerative colitis





# Crohn disease

# Intestinal Complications Of Ulcerative Colitis PERFORATION



Intramural air



Intra- and extramural air



# Intestinal Complications Of Ulcerative Colitis BENIGN STRICTURE





# Computed tomography enterography (CTE)

- The main disadvantage of CTE is radiation exposure.
- Upper gastrointestinal series with small bowel follow-through

# Other modalities Video capsule endoscopy

- For children older than two years
- Risk of symptomatic capsule retention was 5 percent
- 38 percent among those with CD

### **Ultrasonography**

- The sensitivity and specificity 76 and 92 percent ,
- Shows wall thickening and stiffness
- Small intestine contrast ultrasonography;
- oral nonabsorbable anechoic contrast is given by mouth prior to abdominal ultrasonography
- sensitivity of 96 percent and specificity of 100 percent

### **Diagnosis**

► There are no specific diagnostic criteria for IBD.

### **DIFFERENTIAL DIAGNOSIS**

- Rectal bleeding
- Anal fissures or hemorrhoids
- Polyps
- Meckel's diverticulum
- Milk protein-induced proctocolitis

### **Enteric pathogens**

- Salmonella, Shigella, Yersinia, Campylobacter, Aeromonas, enterohemorrhagic
- Escherichia coli, Amoeba, Strongyloides, Trichuris trichiura, and C. difficile
- CMV disease, using sigmoidoscopic biopsies, is suggested for patients with severe colitis that does not respond promptly to intravenous glucocorticoids

- Intussusception
- ► Immunoglobulin A vasculitis
  - ► Henoch Schonlein purpura
- ► Familial Mediterranean fever

#### Growth failure diarrhea

Celiac disease

#### Abdominal pain

- ► Focal abdominal pain, particularly in the right lower quadrant, is more suspicious for IBD (particularly CD).
- differential diagnosis includes appendicitis
- tuberculosis or lymphoma .
- gynecologic disease, vasculitic perforation, or trauma.

# VERY EARLY-ONSET INFLAMMATORY

These children tend to have more severe disease at presentation and are less likely to respond to initial induction therapy

#### Clinical feature

- Young age of onset
- Recurrent infections or unexplained fever Associated features of autoimmunity
- Very severe IBD fistulizing disease or resistance to conventional therapies
- hemophagocytic lymphohistiocytosis (hepatomegaly, fever, cytopenias, high ferritin)
- Lesions of the skin, nails, or hair
- history of cancer
- tissue eosinophilia, villous flattening

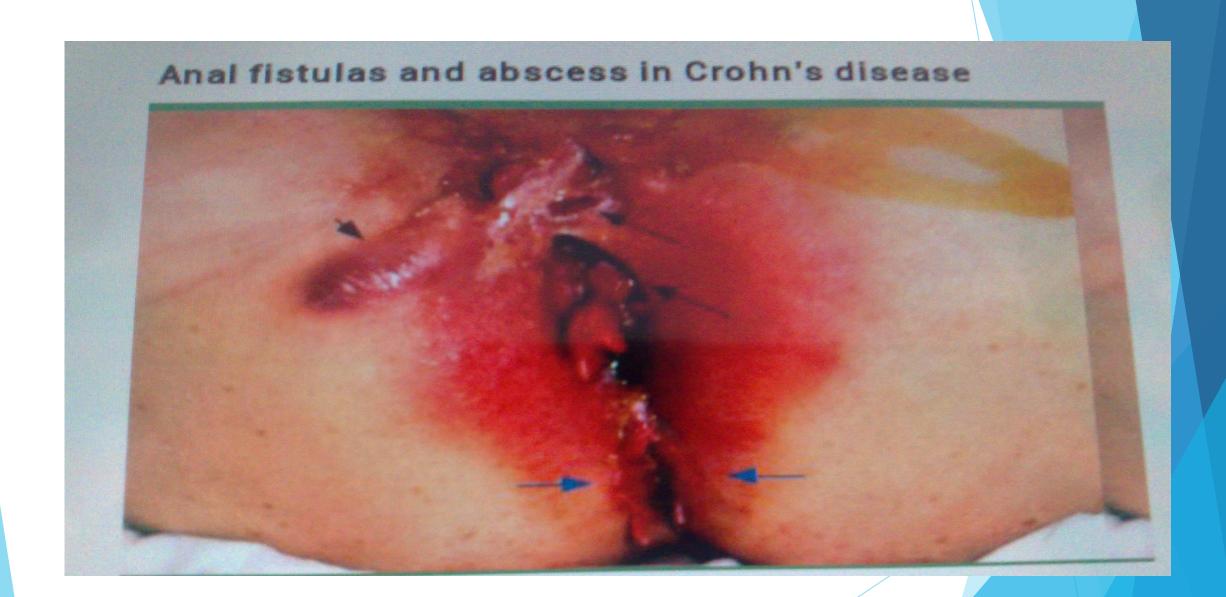
Important causes of monogenic IBD include defects in interleukin-10 (L10) signaling, atypical severe Combined immunodeficiency, common variable immunodeficiency, chronic granulomatous disease and other neutrophil defects, Wiskott-Aldrich syndrome; agammaglobulinemia, hyperimmunoglobulin M syndrome, familial hemophagocytic lymphohistiocytosis, and IPEX.

#### Management approach

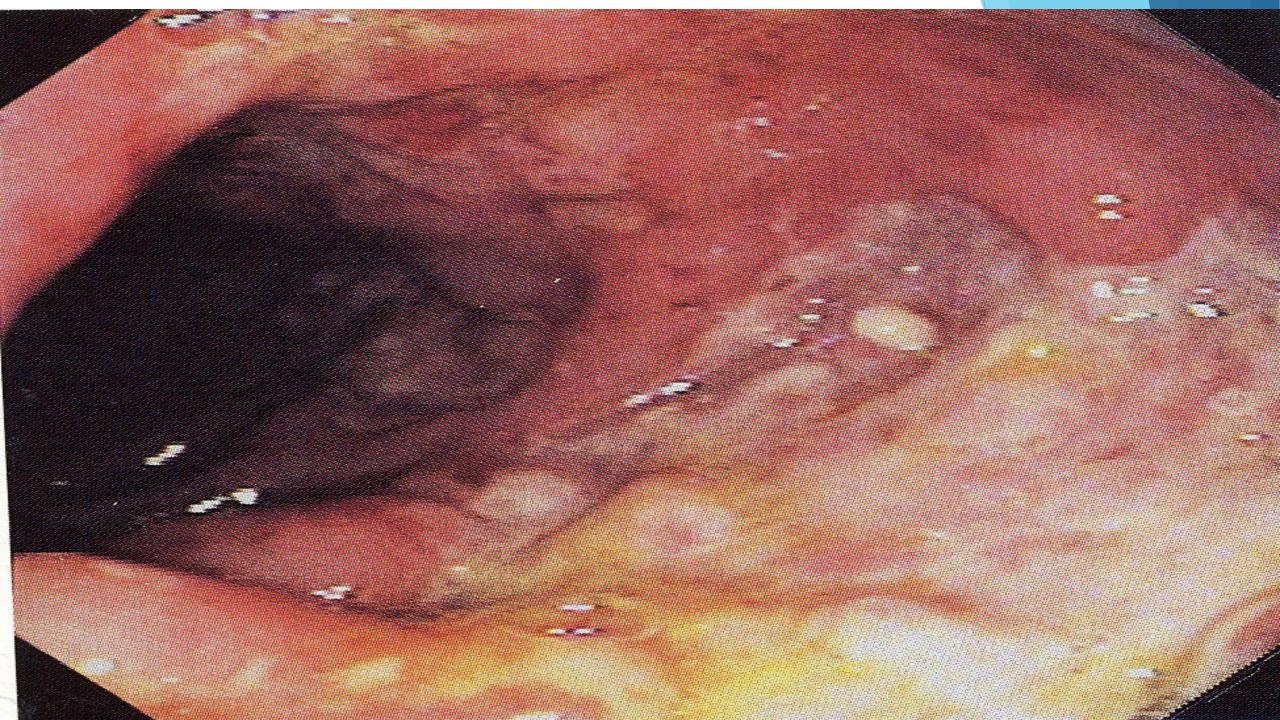
► Hematopoietic stem cell transplantation

### **Features of Crohn disease**

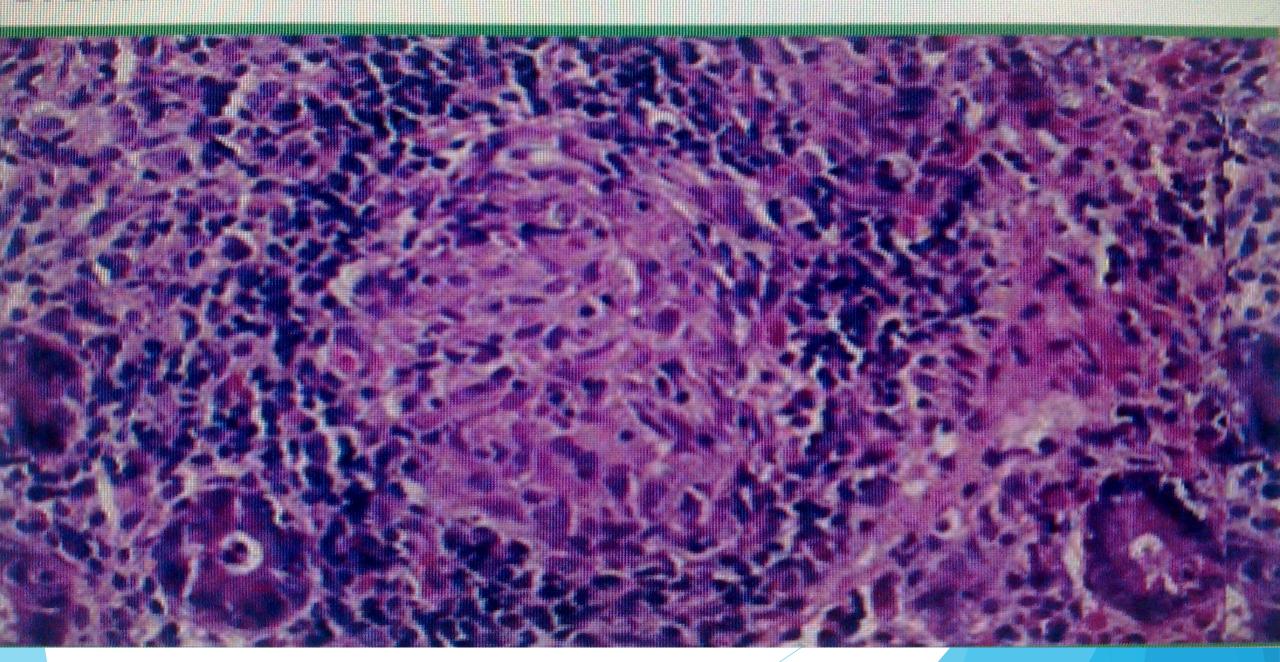
- Physical examination Perianal fistulae, abscess, or large (>5mm) skin tags
- Imaging (MRE) (UGI/SBFT)
- (CTE)
- mucosal ulceration, cobblestoning, narrowing or obstruction, or enteric fistulae
- Gross features at colonoscopy;
- ulceration and stenosis of the ileocecal valve, cobblestoning or linear ulcerations in the ileum, and stricture and fistula formation.
- ► Histopathologic features Noncaseating granulomas 25 to 50 percent







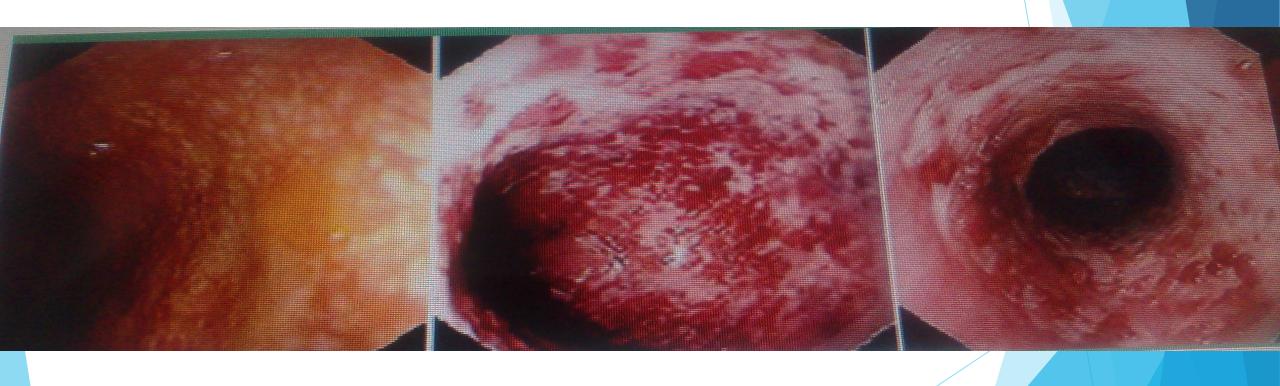
### Crohn disease



### Features of ulcerative colitis

- Continuous process starting at the rectum and extending more proximally into the colon.
- Longer-standing disease may have "pseudopolyps "
- The histopathological features of UC include mucosal and submucosal inflammation, cryptitis, and crypt abscess formation; distortion of the architecture; basal lymphoid aggregates; and the presence of Paneth cells in the left colon.

# UC



# UC



### Non classic features of UC

- Small anal fissures and skin tags; oral ulcers
- Gross or histologic evidence of gastritis
- "Backwash" ileitis
- Periappendiceal inflammation
- Rectal inflammation that is less severe than in the more
- Proximal colon (relative rectal sparing)
- Patchy distribution of colitis

## **Antibody testing**

- ► (P-ANCA)
- ► (ASCA)
- sensitivities (>90 percent in populations with symptoms)
- ASCA antibodies are found in 40 to 80 percent CD, terminal ileum and cecum
- P-ANCA can be detected in 50 to 80 percent ulcerative colitis, 27 percent of adults with CD
- Patients with CD who have P-ANCA antibodies often exhibit UC-like features

► The OmpC is an outer membrane porin, E. coli protein that is immunoreactive to P-ANCA monoclonal antibodies

Antibodies to the bacterial flagellin CBirl are found in approximately 50 percent of individuals with CD and have been associated with small bowel, internal-penetrating, and fibrostenosing patterns.

## Prognosis

## **>UC**

- •†Relapse: younger- female- recurrent relapsers- >70y-Plasmacytosis
- •Colectomy: 5% in first year

20% in 5y

30% after 15-20y 50% → inactive

40% → intermitant

10% → persistant

- •Proximal distribution: 15% in 5 y
- •proctitis: 20% recovery

## prognosis

- >CD
- 1/3 Mild
- 1/3 ex & rem
- •1/3 Chronic active
- •Risk factors: age <40 y- perianal dis- Need to steroids