Basic data

- Name：葉X平
- Age：16
- Gender：Female
- Body Weight：67kg
- Occupation：student
- Date of admission：950522
- Zone of Residency：Taipei

Chief Complaint

- Vomiting and diarrhea for one week

Present Illness

- Vomiting ,watery diarrhea and bloody stool
- Fever and chills
- Whole body artharagia , oral ulcer
- Persistent dull pan-abdominal pain

Physical examination

- General appearance : acute
- Abdomen : soft and mild distention
  diffuse tenderness
  hypoactive bowel sound

Laboratory Data

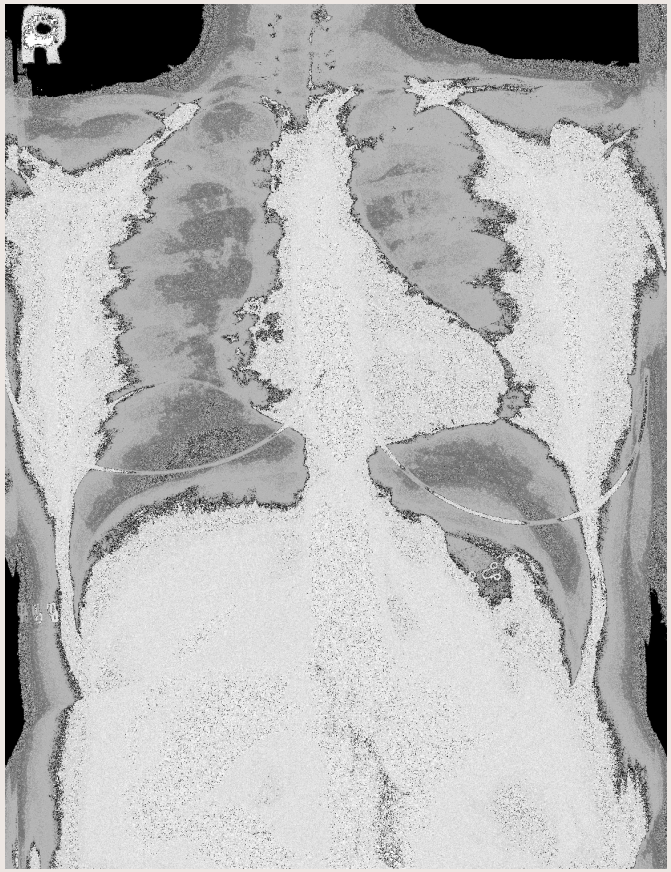
- WBC [4.0-11.0x10.e3/uL] = 21.96
- % Neut [40-74 %] = 84.1
- Amylase [25-125 U/L] = 164
- MCV [80-99 fL] = 77.5
- CRP [0.0-0.8 mg/dl] = 6.20

EKG

- Sinus tachycardia

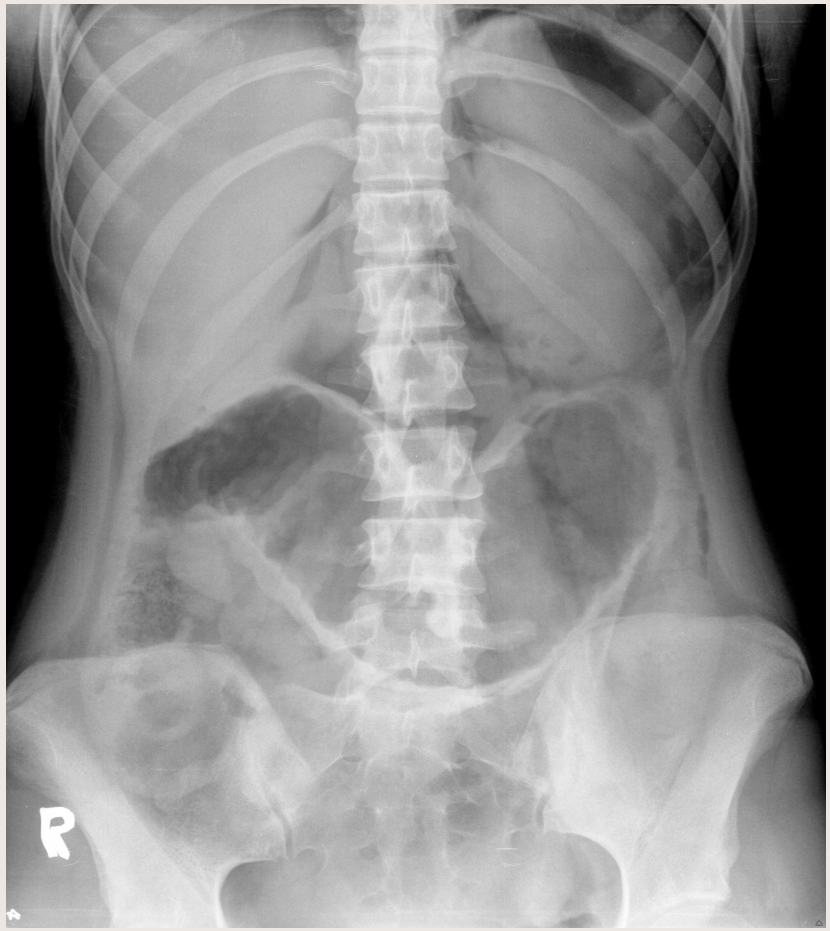
Image

- Pneumoperitoneum
- Subphrenic free gas



Image

- Rigler's sign (double wall)
- Dilated bowel loop
- Subhepatic free gas



Impression

Hollow organ perforation

- R/o Perforated Peptic Ulcer
- R/o Toxic Megacolon
- R/o Acute Colonic Pseudo-obstruction (ACPO)
- R/o Acute Megacolon
- R/o Malignancy

Perforated Peptic Ulcer

- Perforated peptic ulcer -- most common cause of pneumoperitoneum in older children
- Duodenal ulcer perforations are 2-3 times more common than gastric ulcer perforations.
- The incidence of perforation in duodenal ulcer is less then 10%.
- Signs of a large pneumoperitoneum

Toxic megacolon

- nonobstructive colonic dilatation larger than 6 cm and signs of systemic toxicity
- Colitides: inflammatory, ischemic, infectious
- Diagnostic criteria (Jalan et al):

1. radiographic evidence of colonic dilatation
2. any 3 of the following:

   fever (>101.5°F), tachycardia (>120), leukocytosis (>10.5), or anemia
3. any 1 of the following:

   dehydration, altered mental status, electrolyte abnormality, or hypotension.

Toxic megacolon

- Most cases affect young adults, but individuals of any age can be affected
- Distention of the transverse colon associated with mucosal edema

Acute Colonic Pseudo-obstruction (ACPO)

- Ogilvie syndrome
- is a clinical disorder with the signs, symptoms, and radiographic appearance of an acute large bowel obstruction with no evidence of distal colonic obstruction.
- The risk of perforation for ACPO ranges from 3-15% and carries a 50% mortality rate

Acute Colonic Pseudo-obstruction (ACPO)

- Age
  1. generally a disease of elderly patients
  2. may occur in younger patients, particularly those with underlying spinal cord disorders or primary cancer of or metastatic cancer to spinal region

- Clinical presentation:

  Abdominal pain(80%), Nausea and vomiting(80%), Obstipation(40%), Fever(37%), Abdominal distention(90-100%), Abdominal tenderness(64%)

- Imaging Studies:

  Specific attention to the diameter of the colon is important

  If the colonic diameter exceeds 10 cm, decompression of the colon must be considered and expedited.

Acute megacolon

- May be idiopathic, electrolyte abnormality, metabolic abnormality, or certain medications, including anticholinergics, opiates, digitalis, and certain antipsychotic drugs
- can occur in any age group, the typical patient is an elderly person
- spontaneous perforation from nontoxic megacolon is 3%.

Acute megacolon

Clinical presentation

- The abdomen becomes markedly distended, possibly with associated difficulties in breathing.
- No associated peritoneal signs may be present, and all indications for any type of infection may be absent.
- Not having a history of similar episodes of abdominal distension in the past is common.

Imaging Studies:

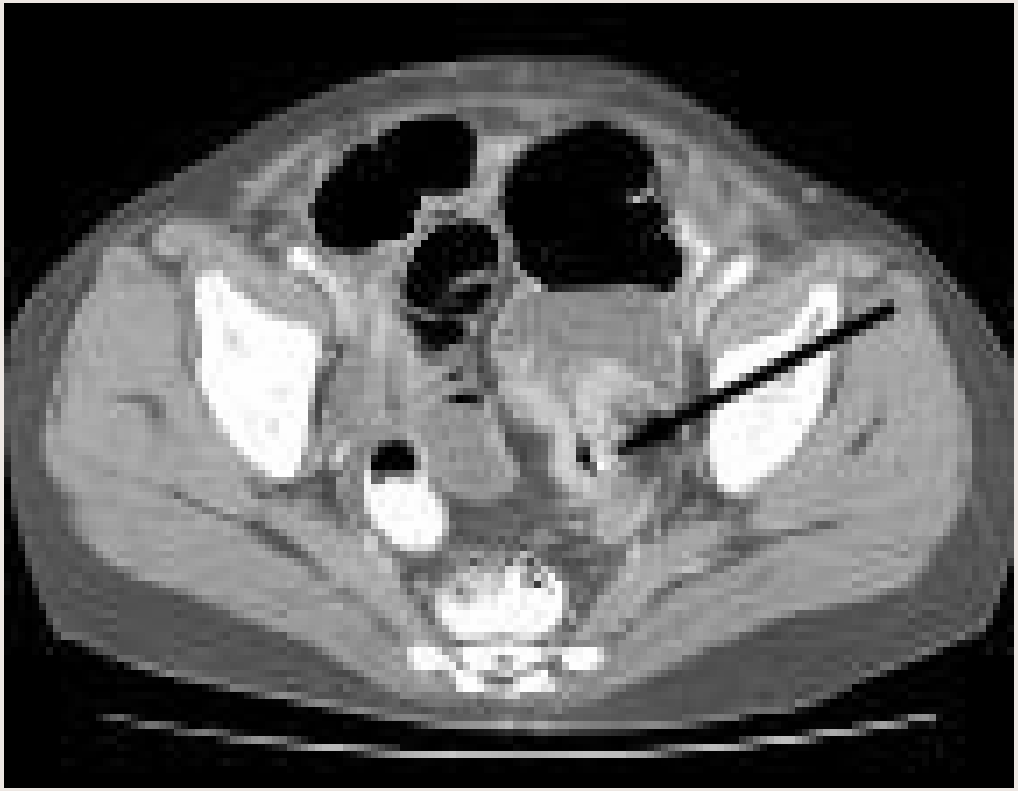
massive gaseous distention of the colon

Malignancy

- Colorectal cancer is rare in children with an incidence of 1.3 to 2 cases per million children
- Most of the cases in children occur in the second decade of life (13-18 y/o)
- abdominal pain, constipation, vomiting, nausea, rectal bleeding, abdomial distension, diarrhea

Malignancy

- CT in a patient with history of caecal carcinoma shows a metachronous annular tumour in the sigmoid causing dilatation of the bowel proximal to it



Plan

- Arrange an emergent operation
- Keep vital sign

OP finding

- Distend abdomen with free air
- About 10cm in length segment of sigmoid colon with inflammatory change. Fibrin coating are found around the area.
- segmental resection of S-colon with Hartmann's procedure and proximal end colestomy
- Resected segment :

  multiple ,various in size ulcers involve entire layer of the colon showed cobblestone appearance.

Post op D/D

Sigmoid Colon Perforation

- R/o Crohn's disease
- R/o viral infection
- R/o bacterial infection

Stool culture

- Salmonella(-)
- Clostridium difficile(-)
- Rota virus(-)

Pathhology(1)

- Specimen:
  - Sigmoid colon, 20cm above the anal verge, segmental resection
  - 11.2 cm. in length and 4.8 - 8.2 cm in circumference
- Gross
  - the serosa is coated by several small patches of fibrinous exduate
  - the intestinal wall in slightly thinner than usual
  - the mucosa surface shows irregular, geographic, and confluent ulcers, predominant arranged along the long axis of the large intestine
  - mucosa is reddening and edematous

Pathhology(2)

- Microscopic

  - multiple ulcers predominantly involving to the submucosa
  - transmural ulcers with bowel perforation accompanied by peritonitis
  - ulcers are broad-based on aggregation of histiocytes and scattered multinucleated giant cells
  - granuloma formation is seen within or near the ulcers

- The features are compatible with Crohn's disease

Final diagnosis

- Crohn's disease

Treatment

- Sulfasalazine

Discussion

Crohn’s disease

- idiopathic, chronic, transmural inflammatory process of the bowel
- can affect any part of the GI tract from the mouth to the anus
- Most cases involve the small bowel, particularly the terminal ileum
- Skip lesions
- Case report in 1976: a 35y/o patient with Crohn's disease involved the entire colon without skip lesions and without terminal ileitis

  (Acta Hepatogastroenterol (Stuttg) 1976 Jun;23(3):227-31)

- Location

  small intestine only (35%)

  small and large intestine (45%)

  large intestine (20%)

- The onset of Crohn disease has a bimodal distribution. The first peak occurs between the ages of 15-30 years; the second, between 60-80 years. However, most cases begin before age 30 years. 25-30% of all patients with CD present when younger than 20 years

Risk factors

- family history (*HLA-DR1* and *DQw5*)
- Smoking
- use of oral contraceptives (2 : 1)
- Diet
- Ethnicity (Jewish)
- female-to-male ratio of 1.1-1.8 : 1

Clinical Presentation(1)

- GI symptoms

  diarrhea, abdominal pain, weight loss,

  rectal bleeding

- may be complicated by intestinal fistulization, obstruction, or both

Clinical Presentation(2)

- Extraintestinal manifestation
  - chronic intermittent fever
  - Iron deficiency anemia
  - arthritis and arthralgia
  - Aphthous ulceration in the mouth
  - erythema nodosum, pyoderma gangrenosum

Clinical Presentation(3)

Perforation rate

- the incidence of colonic and small bowel perforation were 1.6 and 0.7%
- the highest frequency occurring in diseased segments of jejunum (4%)
- Fistula leads to spontaneous intestinal perforation in 1-2% of patients.

Histology

Macroscopic

- deep serpiginous ulcers located transversely and longitudinally over an inflamed mucosa, giving the mucosa a cobblestone appearance
- The lesions are often segmental, being separated by healthy areas.

Microscopic

- Ulcerative inflammation may involve full thickness
- Inflammatory cells invade deep layers and organize into noncaseating granulomas Granulomas (50%)

Lab Data

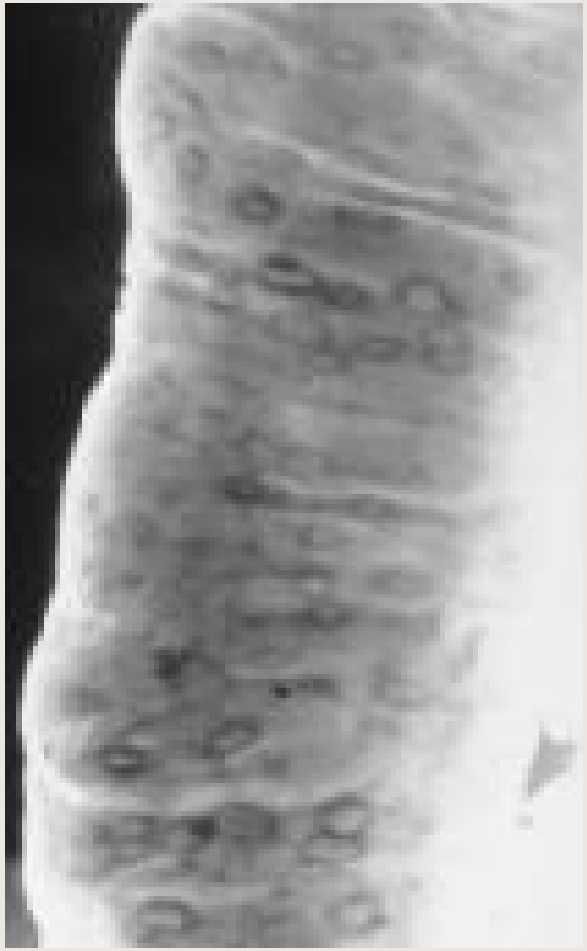
- nonspecific
- microcytic anemia , leukocytosis
- ESR , CRP elevated in 90%
- Hypoalbuminemia, iron, and vitamin deficiencies
- Stool studies
- Anti-*Saccharomyces cerevisiae* antibody (ASCA)
  -- positive in 70% in CD

Image

- skip lesions
- narrowing of the colon

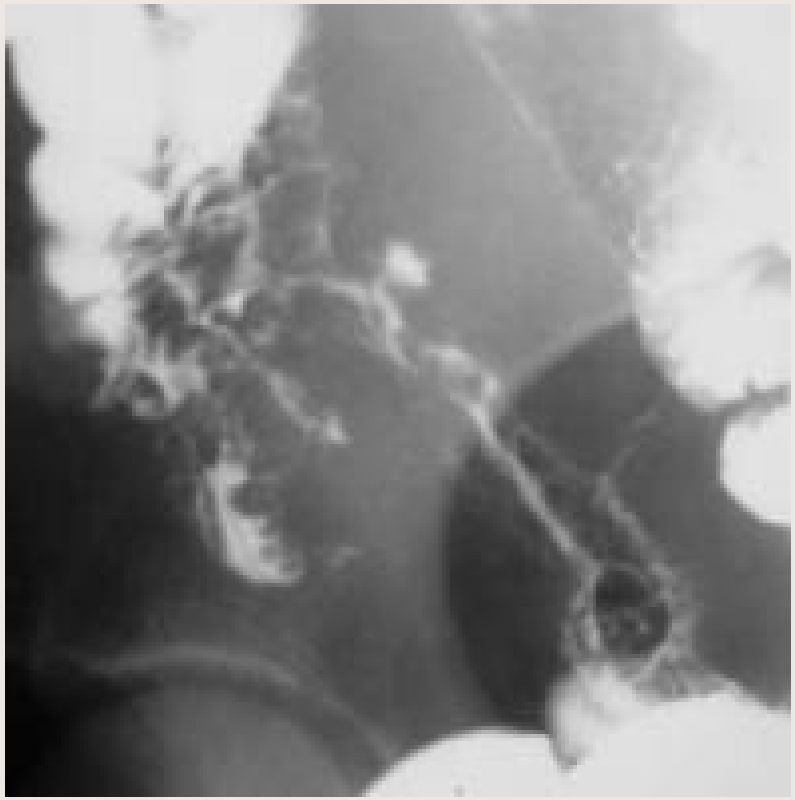
Image

- Double-contrast barium enema examination
- Aphthous ulcers



Image

- narrowing and stricturing
- string sign
- sinus tract

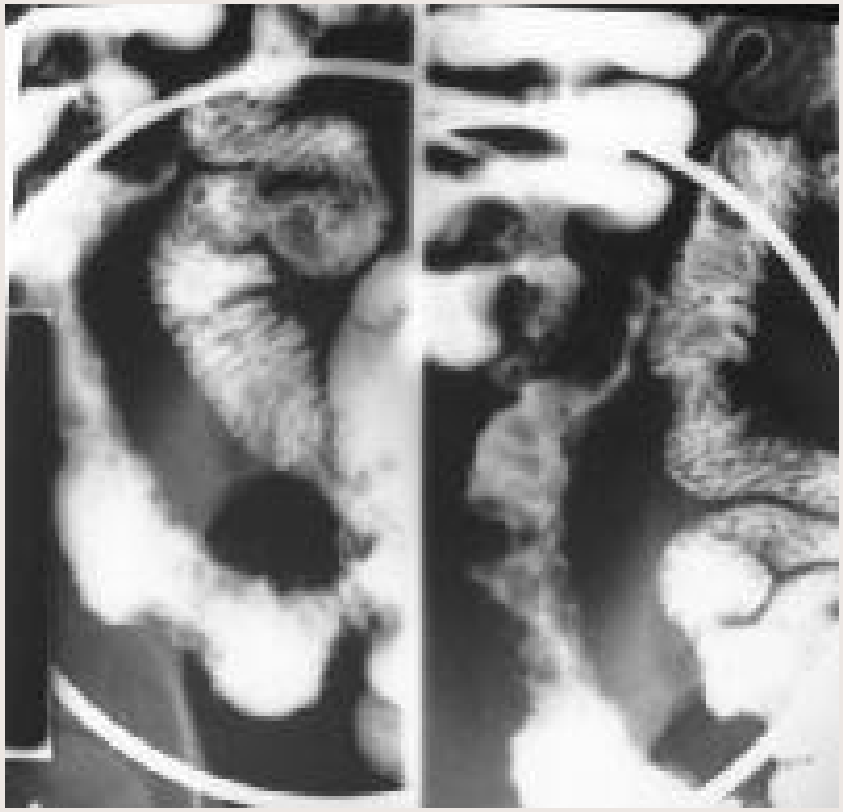


Image

- multiple fistulous tracts between the terminal ileum and the right colon adjacent to the ileocecal valve

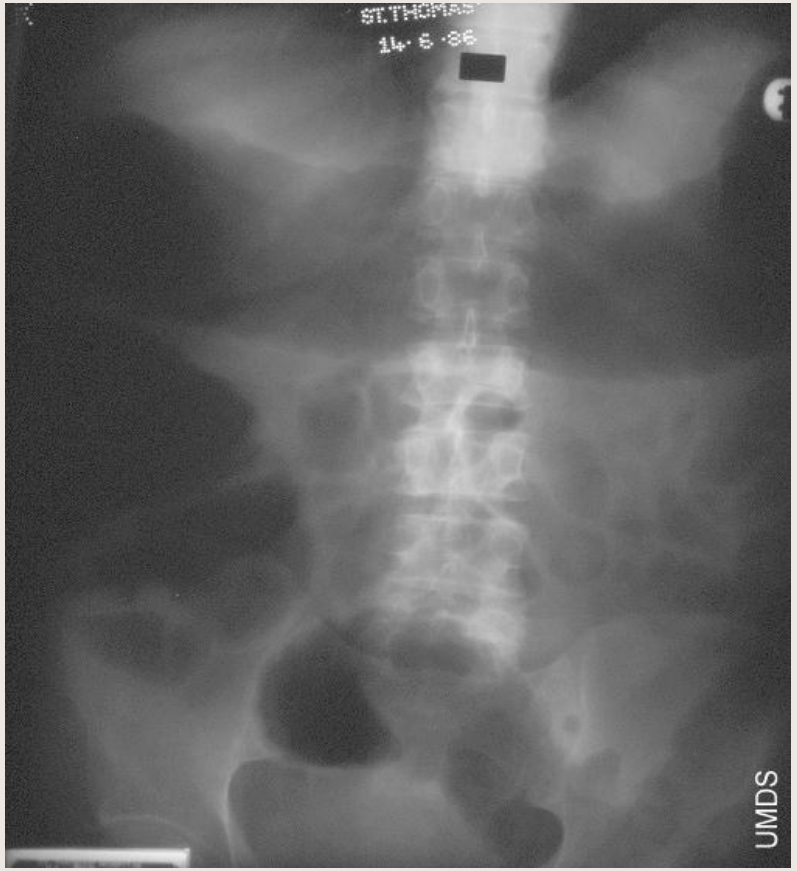
Image

- linear longitudinal and transverse ulcerations
- cobblestone appearance



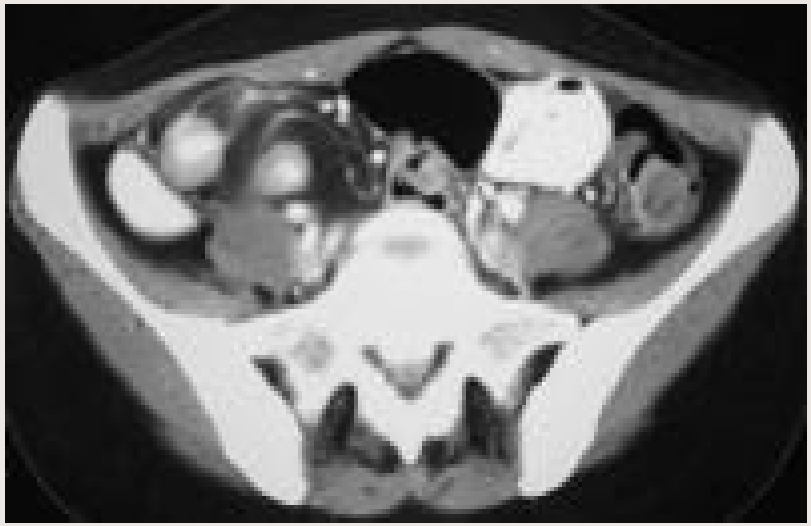
Image

- Crohn's colitis with toxic megacolon
- marked dilatation of the large bowel, from the caecum on the right to a loop of sigmoid seen centrally in the pelvis



Image

- terminal ileal-wall thickening
- adjacent mesenteric inflammatory stranding.



Treatment

- *Anti-inflammatory agents :*
  Sulfasalazine ,Mesalamine
- *Corticosteroids* : ,
  Budesonide
- *Antibiotics* :
  metronidazole and ciprofloxacin
- *Immunosuppressive agents* :
  Azathioprine

Treatment

- Nutritional therapies

  omega-3 fatty acids (fish oil)

- **Surgical Care**

  obstruction, abscess, fistula,

  hemorrhage, perforation

Prognosis

- may have a large impact on the life
- with the appropriate treatment and support, the prognosis is very good
- Colonic malignancy
  - 10-25 years after diagnosis of colitis is estimated to be 8%