Ulcerative Colitis

56

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Ulcerative colitis was first described in 1859 the English physicians, Sir Samuel Wilks who first used this term. While the first pediatric cases were described in 1923 by Helmholz.

56.1 Introduction

There are three diseases within the designation, inflammatory bowel disease (IBD):

- *Crohn's disease (CD)*
- *Ulcerative colitis (UC)*
- *Indeterminate colitis (IC)*

IBD develops at any age and between 10 and 20% are diagnosed in childhood, depending upon the age cut-off used. *Pediatric onset IBD (PIBD)* is more commonly CD while adult IBD is more often UC. However, very early onset IBD (age < 6 years) tends to be more commonly UC again. PIBD tends to be more severe in terms of involvement as well as progression.

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56.2 Epidemiology

There seems a worldwide increase in the incidence of IBD including PIBD and there is also a clear north-south divide with northern states in the USA, Scotland, and northern European countries having a greater incidence than their southern counterparts. The reasons for the increased incidence and pattern of distribution of disease are unclear.

The pathogenesis of IBD is based on a complex interplay of genetics, gut and systemic immune system, the microbiome, and environmental triggers.

56.3 Clinical Features

- Considerable overlap between the presenting features of all types of IBD as well as other disorders such as allergic or infectious gastroenteritis and irritable bowel syndrome.
- Abdominal pain and diarrhea are common features in all IBD but visible blood in stools is more common in UC.
- Weight loss is less prominent in UC compared to CD.

There is often a considerable lag in the diagnosis after symptom onset due to the nonspecific nature of the presenting features.

The differences between UC and CD are outlined in Table 56.1.

Table 56.1 Differences between UC and CD

	Ulcerative colitis	Crohn's disease
Disease extent	Limited to colon, extends from rectum proximally	Occurs anywhere from mouth to anus but rectum is relatively spared
	Perianal involvement is uncommon	Perianal involvement is common
	Stenosis, abscess, and fistulas are rare	Stenosis, fistulas, and abscesses are common
Endoscopic features	Diffuse continuous inflammation extending proximally from the rectum, with friable mucosa, and small superficial ulcers	Patchy inflammation with aphthous or linear ulcers, "*cobblestoning*" of mucosa, active ileitis
	Inflammation limited to mucosa with crypt architectural distortion and crypt abscesses	Transmural inflammation, non-caseating granulomas, rectum sparing, granulomatous inflammation including that of esophagus, stomach, and duodenum
Extraintestinal manifestations	Less common, *pyoderma gangrenous,* and *primary sclerosis cholangitis*	More common, *aphthous stomatitis*, *erythema nodosum*
Surgical treatment	Maybe curative	Reserved for treatment of complications, palliation of symptoms

56.3.1 Investigations

- IBD must be suspected in all children presenting with persistent diarrhea and abdominal pain. Bloody diarrhea is especially a feature of UC.
- Infective causes must be rapidly ruled out by testing stools for common pathogens such as Cytomegalovirus (CMV) and *C. difficile* among others.
- *Fecal calprotectin level* is useful to differentiate between IBD and noninflammatory disease such as irritable bowel syndrome.
- Imaging features are often non-specific. Despite this, initial imaging with US is often used for ease of access and to avoid ionizing radiation. Thickening of the colon with sparing of the small bowel suggests UC and the converse suggest CD.
- *Colonoscopy and biopsies* are the gold standard test for diagnosis (except in acute, severe presentation) and for assessing the extent, and severity of UC. Pediatric-onset UC is more extensive and severe than adult disease, with as many as 90% presenting with pan-colitis.

56.4 Management

The mainstay of treatment for UC as with all IBD, is primarily by medical measures.

- *5-Aminosalicylic acid (5-ASA)* compounds such as mesalamine.
  - First-line treatment for induction and maintenance of remission in UC.
  - Orally or in combination with the rectal route. Rectal administration on its own is reserved for isolated proctitis which is very uncommon in children.
- *Corticosteroids*
  - Second-line treatment for mild-to-moderate UC not responding to 5-ASA.
  - Severe UC is treated with intravenous steroids.
  - Second-generation steroids that are poorly absorbed such as *beclomethasone dipropionate (BDP)* and *budesonide-MMX* may be considered in mild disease refractory to 5-ASA.
  - Not used for maintenance of remission.
  - Thiopurines.
    Used to maintain remission if frequent relapses occur while on 5-ASA.
  - *Infliximab (IFX)*
    Chronically active or steroid-dependent UC that is uncontrolled by 5-ASA and thiopurines.

Unlike CD, UC is amenable to surgical cure and therefore, surgery should be considered early in children and certainly before medical treatment results in irreversible complications.

Indications for Surgery in Children

- Failure of medical treatment.
  - Symptoms despite maximal medical treatment.

  - Growth failure.
  - Adverse impact on education or ability to participate in sport and social activities.
- Emergency surgery for ongoing, severe bleeding, or toxic megacolon.

56.4.1 Surgical Treatment of UC

The primary aim of surgery for ulcerative colitis is removal of the entire colon and rectum. However, simply doing this would result in a *permanent ileostomy* which is not acceptable to most and therefore, various operations have been devised to restore bowel continuity. Restoration of bowel continuity can be done at the same time as the *proctocolectomy* or as a separate procedure, in which case the rectum is initially left in situ as a Hartmann[1] pouch. This type of operation is the procedure of choice in an emergency situation.

As the aim is to retain as little rectal mucosa as possible, an *Ileo-rectal anastomosis* for restoration of bowel continuity is no longer considered acceptable. The rectum is completely excised or a *rectal mucosectomy (Soave-type procedure)* is performed. Rectal mucosectomy has the advantage of a reduced risk of pelvic nerve and urinary tract damage.

- The ileum is anastomosed just above the transitional epithelium of the anal canal.
- A straight ileoanal anastomosis may be performed or the ileum may be folded on itself in various configurations to increase reservoir capacity, the most commonly performed such procedure being an *ileal J-pouch*. This is often considered the procedure of choice for restoration of bowel continuity after total proctocolectomy.
  - The main advantage of a J-pouch over a straight ileoanal anastomosis is that it results in a ↓stool frequency.
  - ↑ complication rate and incidence of pouchitis.
  - The anastomosis may be hand-sewn or stapled, though the use of a circular stapler may be limited by the size of the child.
  - An ileostomy is often created to divert feces and allow the anastomoses to heal though a single-stage operation is also feasible.

Minimally invasive techniques are gaining traction worldwide and a laparoscopic proctocolectomy and ileal pouch anastomosis (IPAA) can be done with good outcomes. A laparoscopy-assisted procedure is also possible in which the colon mobilization is done laparoscopically leaving the removal of the colon and creation of the ileal pouch to be done via a small lower abdominal or Pfannenstiel incision.

[1] Henri Albert Hartmann (1860–1952)—French surgeon working in the Hôtel-Dieu in Paris described this in 1923.

56.4.2 Complications

Ileal pouch-anal anastomosis is associated with a significant complication rate of 55–75% depending upon the length of follow-up. Up to 50% of patients will require reoperation. The most commonly reported complications are:

- Wound complications
  - Infection, dehiscence (5–20%)
- Anastomotic leak (0–15%)
- Anastomotic stricture (10–25%)
- Fistula, especially pouch vaginal fistula (3–30%)
- Small bowel obstruction (15–30%)
- Pouchitis (30–70%)
- Recurrent pouchitis (5–35%)
- Pouch failure is defined as excision of pouch and permanent ileostomy (up to 10%)
- Afferent limb syndrome (obstruction of the small bowel at the pelvic brim)
- SMA syndrome
- Functional complications
  - Incontinence (up to 15% daytime and 30% at night)
  - Infertility and sexual dysfunction

- *Nonspecific inflammation of the pouch (pouchitis)*
  This is the most common complication of Ileal-pouch surgery and manifests as abdominal pain, cramps, urgency, frequency, tenesmus, and hematochezia.[2] Pouchitis is diagnosed based on clinical features. Investigations are not required before treatment. It is treated with a course of antibiotics, usually metronidazole and ciprofloxacin. Endoscopy and biopsy are reserved for recurrent or refractory pouchitis to rule out anatomical problems such as anastomotic stricture. Biopsy is done to look for CD in the pouch. This is an important cause of pouch failure alongside anastomotic leak, prolonged disease, and treatment with biologic agents prior to pouch formation.
- *Acute Severe Ulcerative Colitis (ASUC) and Toxic Megacolon*
  Up to a third of pediatric patients with UC will present acutely and require hospitalization within the first 3 years of diagnosis. This acute, severe form of UC is diagnosed based upon clinical criteria including frequency and consistency of stools, blood in stools, abdominal pain, blood in stools, and limitation of activity *(Pediatric UC Activity Index—PUCAI).* Intravenous steroid administration is the mainstay of treatment. Infective causes such as *Clostridium difficile* and CMV should be treated as required. Prophylactic antibiotic therapy is not useful and neither is "gut-rest." Second-line treatment includes Infliximab.

[2] Passage of blood with stools. (Greek) αἷμα—blood and χέζειν—defaecate.

- *Toxic megacolon*
  This is a form of acute severe UC in which the colon is dilated (>5.6 cm, or >4 cm if the child is <11 years old) in the absence of mechanical obstruction and there are features of systemic sepsis. The colonic dilation may be segmental or generalized. Colonic perforation is a feared complication, the clinical features of which may be masked by steroid administration. Serial abdominal radiography is therefore used until the patient's clinical condition improves. Treatment is based upon fluid resuscitation and correction of electrolyte imbalance as well as broad-spectrum antibiotic cover.
  Failure of ASUC to respond to medical therapy, colonic perforation, sepsis and bleeding are indications for emergency surgery, though it is becoming less likely than, the past. As mentioned previously, subtotal colectomy with Hartmann pouch (blind rectal stump) and end ileostomy is the procedure of choice and this can be safely achieved laparoscopically even in the acute situation.

Further Reading

Goldstein AM, editor. Inflammatory bowel disease in children. Semin Pediatr Surg. 2017;26:343–404.