



Bruce Jaffray

Burrill Bernard Crohn (1884–1983)

BBC was a gastroenterologist working in Mount Sinai Hospital, New York. He published 14 cases of “regional ileitis” in 1932 with Ginzburg and Oppenheimer in the JAMA.

55.1 Epidemiology

The incidence of Crohn's disease (CD) in the West has undergone a steady increase over the past 30 years, with some evidence of a recent plateau. A similar pattern but with a time lag is present in developing countries.

- 6.4/100,000 children per annum—Crohn's disease
 - Prevalence 25/100,000 children
- 2.35/100,000 for ulcerative colitis
- 10–15% of inflammatory bowel disease (IBD) cases present in childhood
 - Tendency for younger children to present with colitis.
- M = F
- Marked racial differences in the incidence of the disease.

Children with presentation <6 years of age are characterized as very early-onset IBD [5], for whom there may be an association with antibiotic exposure during pregnancy.

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55.2 Etiology

Crohn's disease results from an interaction between genetic susceptibility, the host's immune system, environmental factors, and the gut microbiota. Chief among causative factors which can be altered is tobacco use; early smoking doubles the odds of developing CD.¹ The importance of environmental factors is illustrated by the increased incidence of CD among the first-generation children of immigrants to Western societies.

55.3 Pathology

- Macroscopic
 - Ileum—often segmental distribution with cobblestone ulceration, fistulae and bowel wall thickening, fat wrapping, and strictures.
 - Colon but typically with rectal sparing.
- Microscopic
 - Discontinuous chronic inflammation, transmural involvement, serositis, lymphoid aggregates, granulomas, and muscular hypertrophy.

55.4 Clinical Features

There is no typical presentation of CD in children.

- 5% of patients have a first-degree relative with IBD.
- Abdominal pain is common, but the vast majority of children with chronic abdominal pain do not have CD. However, abdominal pain provoked by eating is often seen with ileal CD.
- Cessation of growth
 - A child who was previously growing normally may cease growth as a first manifestation. Inspection of a centile chart with the current weight and the weight 1 year previously can be highly suggestive.
- Perianal pathology (Fig. 55.1)
 - Abscess or fistula. An edematous perianal skin tag is a tell-tale sign.
- Diarrhea and bloody stools
 - May indicate colitis, and are more commonly seen in younger children. It is an error to assume that a child without bloody stools cannot have Crohn's disease.
- Intestinal obstruction, or internal fistulation are late complications of the disease, and would be unusual as presenting features where there is access to advanced healthcare.

¹By contrast—cigarette smoke appears to protect against the development of ulcerative colitis.

Fig. 55.1 Crohn's disease of perineum



Extraintestinal manifestations include:

- Joints
 - Peripheral arthritis, axial arthropathy
- Skin
 - Pyoderma gangrenosum
 - Erythema nodosum
 - Psoriasis
- Eyes
 - Uveitis
- Mouth
 - Aphthous stomatitis

While typically developing years after the gut presentation of the disease, these sites can occasionally be the first manifestation of the disease.

55.5 Differential Diagnosis

The typical presentation is with terminal ileal disease. In the West, abdominal tuberculosis as a cause of a small bowel stricture or fistula is extremely unlikely, but maybe a valid concern where tuberculosis remains prevalent [11]. Very rarely *Yersinia* infection can be a cause of granulomatous change in the appendix [12].

An issue much discussed, but seldom encountered, is the child operated upon for acute appendicitis in whom CD suspected. The appendix should be removed and subjected to histological examination. The ileal disease should be left alone, unless the cause of a complication such as fistulation, which would mandate surgery. The concept of appendectomy leading to fistulation in the presence of CD has yet to occur in the author's practice.

If the presentation is with colitis, then there may be considerable difficulty in distinguishing CD from UC, even after biopsy. As a practical issue for surgeons, the distinction is immaterial since urgent surgery for both conditions will consist of a subtotal colectomy with ileostomy. Elective surgery will follow diagnostic workup by gastroenterologists, but it is accepted that a proportion of patients thought to have ulcerative colitis will subsequently have their diagnosis revised to Crohn's disease, and vice versa.

55.6 Investigations

- *Fecal calprotectin.*
 - 90% sensitivity and 75% specificity for the diagnosis of IBD in children [13].
 - It is also useful for the assessment of mucosal healing in response to therapy [14].

Measurement of other surrogates of inflammation such as a full blood count, serum albumin, or C-reactive protein are useful screening adjuncts, but fecal calprotectin outperforms all other tests as a first choice investigation.

Coupled with measurement of fecal calprotectin, abdominal ultrasound should be used as a screening tool for CD in children. The possibility of a child having IBD in the presence of a normal fecal calprotectin and abdominal ultrasound approaches zero.

Other imaging modalities such as MRI enteroscopy, capsule enteroscopy, and CT scanning are used, but the need for these modalities is small once fecal calprotectin is combined with abdominal ultrasound.

If screening assessments suggest the presence of inflammatory bowel disease then combined upper and lower gastrointestinal endoscopy with ileoscopy and biopsy allows determination of disease type and extent. Ileal histology is the gold standard for the diagnosis of Crohn's disease in children.

55.7 Management

There has been a paradigm shift from the control of symptoms to mucosal healing.

Treatment with *anti-TNF therapy* (e.g., infliximab and adalimumab) based on the objective assessment of mucosal healing is now the accepted modality. However, there is uncertainty whether the use of anti-TNF therapy has led to the hoped-for reductions in surgical resections for CD.

Optimal outcomes result from a multidisciplinary approach with close collaboration between pediatric gastroenterology and specialist pediatric surgeons. Surgery is indicated for the failure of medical therapy to induce remission or where there are complications of the disease such as obstruction or internal fistulation. A more complex issue is whether children with isolated terminal ileal thickening are better served by localized resection or medical therapy as first-line therapy.

Surgery in Crohn's disease cannot cure the disease—in contrast to ulcerative colitis.

55.7.1 Indications for Surgery

- First-line therapy for isolated ileal disease.
- Failure to achieve mucosal healing with medical therapy Complications, e.g., internal fistulation or perforation.
- Perianal manifestations.

Surgery for childhood CD has changed profoundly as a result of improvements in medical therapy. Severe disease in children rendered Cushingoid by prolonged steroid therapy should no longer be seen. The typical presentation now is with a short ileal segment that is resistant to attempts at mucosal healing; there is often little macroscopic disease to guide the surgeon, and resection is dictated by preoperative imaging. The incidence of colectomy for Crohn's colitis has also markedly declined.

55.7.2 Surgical Options

For the typical case, *minimally invasive ileocecal resection* should be the gold standard. The principal advantages are minimization of peritoneal adhesion formation with resulting decrease in risk of adhesional obstruction, reduction in postoperative pain, leading to early mobilization, and improved cosmesis. Practical issues are the siting of the specimen extraction wound. Use of a Pfannenstiel incision leads to excellent cosmesis, but it is difficult to mobilize the colon to reach the incision to allow an extracorporeal anastomosis. The alternative of intracorporeal anastomosis increases the technical difficulty of the procedure considerably. Use of a minilaparotomy midline wound may be a more practical alternative for most surgeons, allowing laparoscopic mobilization and division of the mesentery, with a technically straightforward extracorporeal anastomosis.

There is current research interest in the possibility that wide mesenteric excision may reduce the risk of disease recurrence [22].

There is controversy over the management of *segmental Crohn's colitis*. While segmental resection is possible, there is a significant risk of anastomotic recurrence leading to a need for a further resection. However, it is possible to perform segmental resection with either no further surgery, or further surgery postponed for several years. Many teenagers will want to take the risk of further resection in preference to a permanent ileostomy.

Urgent surgery for colitis consists of *subtotal colectomy with ileostomy*. The rectum should be stapled closed at the peritoneal reflection. Use of a distal mucous fistula in this situation is unnecessary.

If there is rectal sparing and no severe perianal disease, then ileorectal anastomosis is possible. However, in the author's experience the combination of severe left-sided colitis requiring resection coupled with a normal rectum is a rare phenotype.

Complex complications are now rarely seen, since children should be put forward for resection before their development. However, the following may be encountered.

Internal fistulation may be between adjacent loops of small bowel, or between the ileum and the sigmoid colon, or between ileum and bladder or vagina. Management consists of disconnection of the fistula, with resection of bowel back to macroscopically disease-free margins. Adherence of the ileum to the adjacent sigmoid may suggest the need for ileal and sigmoid resection, however, in the author's experience, the sigmoid is "an innocent bystander," and ileal resection alone is enough.

Historically, multifocal small bowel CD was often a cause of short bowel syndrome. The need to avoid excess bowel resection led to the use of stricturoplasty to deal with multiple strictures. The changing phenotype of Crohn's disease has made this a very rare procedure.

55.7.2.1 Fistulas

External fistulation following an anastomosis may be encountered. An apparently successful procedure is complicated by the discharge of bowel content through the wound. In a stable child without signs of peritonitis or sepsis, conservative management can be tried. Such fistulas can follow a surprisingly benign course, closing spontaneously in days or weeks. However, if there are signs of sepsis, or failure to close, the anastomosis will require resection and temporary exteriorization.

55.7.2.2 Perianal disease

Perianal disease is frequent, painful, and distressing. Perianal abscesses should be drained as a matter of urgency. Simple drainage is sufficient; packing is unnecessary and painful. Not all abscesses are clinically evident. Drainage may be dictated by MRI appearances, and the author has encountered abscesses that are neither visible nor palpable, but incision over the site indicated by MRI has led to release of pus.

Laying open of fistula-in-ano should be avoided. Too often this leads to a large indolent ulcer that remains unhealed 6 months later.

Placement of *setons*² is a helpful adjunct while awaiting for the effects of anti-TNF therapy. It is frequently the case that the external orifice of the fistula can be cannulated, but the probe cannot be made to reach the lumen of the ano-rectum. With one finger within the rectum, repeated gentle passage of the probe usually lead to a point where there is only thin mucosa separating the probe from the finger. It is a mistake to think that by completing the passage of the probe one is creating a fistula; the internal orifice is already there, but tissue edema prevents the probe from reaching the lumen. The probe should be passed and a seton inserted. These should be left loose and not tightened.

²Seton—derived from Latin *seta*—a stiff hair or bristle.

Practice Point

Crohn's disease is a pro-thrombotic condition, and all intestinal resections should be performed with deep venous thrombosis prophylaxis, even in children.

Ano-rectal strictures can be treated by serial dilatation under anesthetic, again while waiting for the effects of medical therapy.

Very rarely, the complex ano-rectal disease may require temporary diversion with a proximal stoma for a period. Reversal of the stoma a year later does not invariably lead to disease flaring again.

55.8 Outcome

- Early complications include anastomotic leak, hemorrhage, and external fistulation.
- Late complications include disease recurrence, typically peri-anastomotic.

Further Reading

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