THE CLINICAL PRESENTATION AND PROGRESS OF INFLAMMATORY BOWEL DISEASE

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Does inflammatory bowel disease present differently or behave unusually in the young adult? There are some important differences, these occur particularly in children with Crohn's disease around the age of puberty. Such variations may therefore be seen in young service personnel.

Clinical presentations

1. *Proctocolitis*. These patients usually present with bloody diarrhoea (the problems of infective causes has already been touched upon). A few present with rectal bleeding but without diarrhoea, these are mostly patients with proctitis. Full examination, sigmoidoscopy and biopsy and barium enema will be required. Patients with anal lesions, a normal rectum or localised lesions with ulceration are classified as likely to have Crohn's disease. Those with no anal lesion, a diffusely granular rectum and either a normal or diffusely abnormal sigmoid or colon on barium enema are called idiopathic proctocolitis. The problem that must be emphasised is that this second group cannot be definitely diagnosed without rectal biopsy and follow up. The presence of a single granuloma is enough to change the diagnosis to Crohn's disease, follow up may also change the diagnosis and occasionally examination of a colectomy specimen may alter the diagnosis. Hence the need for rectal biopsy at every stage of the disease.

2. *"Classical" Crohn's disease*. A trilogy of abdominal pain, diarrhoea and weight loss is normally present. If the patient has an abdominal mass and an anal lesion the diagnosis can be made with some confidence. So the recognition of anal lesions is all important, particularly as many are surprisingly pain-free so that the patient describes them as piles or is unaware of their presence.

Crohn's anal lesions are either a fissure or a fistula, occasionally a diffuse inflammation and oedema of the anus is the sole finding. The lesions have a characteristic cyanotic colour but as with the rectal findings should always be confirmed by histology if the diagnosis has not been made.

Intestinal disease is often accompanied by (or may occasionally present with) malabsorption, chronic obstruction, internal and external fistulae and protein losing enteropathy. A discussion of all these complications is beyond the scope of this presentation.

3. *Duodenal Crohn's disease*. It is now well recognised that Crohn's disease may be a widespread abnormality in the small and large intestine even though the obvious manifestations are local. Duodenal Crohn's disease is yet another local lesion, this may present with dyspepsia and radiological and endoscopic abnormalities and is sometimes misdiagnosed as duodenal ulcer. However systemic illness and anaemia should alert the clinician. Furthermore the endoscopic appearance may be more diffuse than simple ulceration with a cobblestone mucosal change and multiple ulcers.

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4. *Pyrexia of unknown origin.* Crohn's disease occasionally presents with fever and no bowel symptoms, this can occur at any age but is more common in the young.

5. *Anorexia nervosa.* This is an important differential diagnosis in the young adult, particularly if the patient is female. Anorexia is commonly present in inflammatory bowel disease at all ages but in the young woman wasting and amenorrhoea may closely mimic anorexia nervosa. Signs of systemic disease, fever, anaemia and a raised sedimentation rate should arouse suspicion.

6. *Failure to grow.* In children physical development may be retarded and normal sexual maturation may be delayed in patients about the years of puberty. This may bring these cases to the endocrinology department.

7. *Unexplained anaemia.* Iron deficiency, resumably due to gastro-intestinal bleeding, may antedate the usual symptoms by a considerable period. In my experience this sometimes happens at puberty.

Progress

Once the diagnosis of idiopathic procto-colitis or Crohn's disease has been established a pattern of extension and relapse can be predicted. This prognosis can be more accurately given in procto-colitis (Powell-Tuck *et al*¹, Ritchie *et al*²). This work from St Mark's Hospital is summarised in Tables I and II. Further-

Table I
Prognosis of proctitis
(From Powell-Tuck *et al* 1977)

Extension to descending colon	Extension to hepatic flexure
5 per cent at 5 years	4 per cent at 5 years
12 per cent at 10 years	6 per cent at 10 years

Table II
Prognosis of colitis
(From Ritchie *et al* 1978)

Extension of distal colitis	Extension of substantial colitis
18 per cent at 5 years	21 per cent at 5 years

more the mortality of the disease is examined in this series; remarkably there were no excess deaths due to colitis in 269 patients. This is a great achievement and reflects both the advances in therapy and the excellence of the centre.

The course of Crohn's disease is notoriously more difficult to classify and prognose mainly because the manifestations are so diverse. The high recurrence rate after surgical treatment is well-known and will be discussed in a later section. The natural history of Crohn's disease is well described by Truelove and Pena³, extension of the lesions is common. An attempt has been made to correlate the pattern of the disease and its complications with the site of the presenting lesion (Farmer *et al*⁴) (Table III). It can be seen that arthritis is strongly associated with

Table III
Patterns of Crohn's disease
(From Farmer et al 1975)

	Small intestine (percentage)	Ileocolic (percentage)	Colonic (percentage)
Cutaneous and internal fistulae	17	34	16
Obstruction	35	44	17
Anal disease	14	38	36
Arthritis	4	3.6	16

colonic disease as has been emphasized by Dr Jewell. British experience would put a higher figure on anal disease particularly where the colon and rectum are involved, about 75 per cent of these patients have anal lesions in our experience.

A recent series compares the patterns of Crohn's disease in childhood (some of these patients were up to nineteen years old) (Gryboski and Spiro⁵). This shows that colonic disease is uncommon in this age group, thus confirming the opinion that colonic Crohns is more a disease of older people (Table IV).

Table IV
Pattern of Crohn's disease
(From Gryboski and Spiro 1978 and Farmer et al 1975)

	Adult (percentage)	Children (percentage)
Diffuse small intestine Ileal	28.6	20 20
Ileocolic	41	52
Colonic	27	8
Anorectal	3.4	

Mortality in Crohn's disease remains substantial, a 21.4 per cent cumulative fatality rate is given by Truelove and Pena. When actuarial analysis is used there is a steady widening of the difference between the expected and observed deaths over fifteen years. This highlights a difference between colitis and Crohn's disease, in colitis most deaths used to occur in the first year while in Crohn's disease the danger increases as time goes by.

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