THE ROLE OF SURGERY IN INFLAMMATORY BOWEL DISEASE:

SIX CASE HISTORIES

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INTRODUCTION

In 1878, Weir Mitchell and Alexandria, the first successful treatment of gout was performed. This was a significant breakthrough in the treatment of gout, which had previously been difficult to manage. Mitchell's innovative techniques involved the use of a new drug, which he had synthesized, to help control the symptoms of gout.

The inflammatory process associated with gout is still the subject of ongoing research, but as yet no definitive evidence exists to support Mitchell's findings or those of any other physician who has been associated with gout.

This inevitably leads us to question the role of the management of patients afflicted by these disorders and the relative roles of the physician and the surgeon in treating such conditions.

Whilst on a student attachment with a general practitioner, I was able to experience something of the scope of surgery and the treatment of inflammatory joint disease. I was introduced to the care of what I came to know as 'knee disease' and to various surgical procedures. I was also able to see the subtleties and complications that can occur.

INTRODUCTION
In 1875 Wilks and Moxon first described the disease entity known as ulcerative colitis. Thirty-eight years later Dalziel described chronic interstitial enteritis and the disease - regional ileitis - was later defined by Crohn in 1932.

The inflammatory bowel diseases have been the subject of considerable research but as yet little hard evidence as to their aetiologies has been unearthed. This inevitably leads to great debate as to the management of patients afflicted by these disorders and the relative roles of the physician and the surgeon are highly controversial.

Whilst on a student attachment with a general surgical unit I was able to experience something of the place of surgery in the management and treatment of inflammatory bowel disease. I have selected six patients in the care of whom I have been involved to illustrate some of the areas of surgical involvement in these diseases and some of the dilemmas and complications that may arise.
CASE ONE

Mr. D. H. C. (Staff 21466)

Mr. D. H. C. was admitted to hospital in May 1936 as the case of tuberculous meningitis. He had no history of contact with any other person suffering from tuberculosis. His general health had been average and he had never been ill. He was a farmer by occupation. He had no history of any surgical procedures.

On this treatment regime, however, he continued to suffer recurrent sino-pulmonary infections, developing bronchiectasis and recurrent endocarditis and staphylococcal pneumonia. In 1950 because of consistently low serum immunoglobulin, therapy was changed to fresh frozen plasma. In 1964 he was investigated for weight loss, abdominal pain, and neurological alteration. No cause for the malabsorption was discovered and he was successfully treated with a combination of antacid and antimicrobial agents as an alternative to intravenous immunoglobulin.

CASE ONE

In October 1965 he was again admitted after he developed painful lesions in the mouth with associated pyrexia. These settled with a combination of antibiotics and high dose intravenous immunoglobulin. Radiographic and follow through of this time displayed acute extensive small bowel obstruction consistent with mild strictureing, obstruction and diverticulosis.

Although various surgical procedures were discussed with the patient, none were carried out due to his general condition. During his hospitalization, he began to exhibit signs of neurological involvement, particularly weakness and tetraparesis.

The patient died in hospital with no apparent immediately pathological cause of death.
CASE ONE

MR D. McN (30.05.64)

Mr McN has suffered recurrent infections since the age of three months and at 30 months a diagnosis of Bruton type X-linked hypogammaglobulinaemia was made. A younger brother also has the condition. In 1974, after extensive investigation of his immunological status, he was commenced on intramuscular gammaglobulin injections at a dose of 0.25g/Kg/week.

On this treatment regime, however, he continued to suffer recurrent sino-pulmonary infections, developing bronchiectasis and recurrent sinusitis and otitis media. In 1980 because of persistently low serum immunoglobulin, therapy was changed to fresh frozen plasma. In 1981 he was investigated for weight loss, abdominal pain, anorexia and oral ulceration. No cause for his malabsorption was discovered and he was successfully treated with a combination of metronidazole, cotrimoxazole and an alteration of his immunoglobulin replacement therapy to intravenous immunoglobulin.

In October 1982 he was again admitted after he developed painful lesions in the mouth with associated pyrexia. These settled with a combination of antibiotics and high dose intravenous immunoglobulin. Barium meal and follow through of this time displayed quite extensive small bowel abnormalities consisting of mild stricturing, ulceration and diverticulosis.

Although suffering regular chest infections he had no diarrhoea or any other symptoms referable to his abdomen until he presented in April 1985. He gave a four week history of increasing colicky upper abdominal pain, anorexia, a four kilogram weight loss and diarrhoea associated with the passage of mucus and blood. On examination the only positive finding was the he appeared grossly underweight. (Platelet count
investigations showed severe iron deficiency anaemia with haemoglobin of 9.1g/dl, an Erythrocyte Sedimentation Rate of 12 millimetres in the first hour and a reduced albumin of 30g/L. A bone marrow examination revealed no evidence of lymphoma. Microbiological examination of stools showed no enteric pathogens and xylose absorption test was within normal limits. His serum gammaglobulin level was unmeasurable despite being on high replacement therapy. On endoscopy the stomach appeared normal but the second part of the duodenum was grossly abnormal with hypertrophy and disruption of mucosal folds associated with ulceration. These appearances were thought to be consistent with lymphoma but histological examination of biopsies taken were non-specific. A barium meal and follow through was performed which was reported as showing ulceration, tubular narrowing, saculation and thickening of bowel wall together with effacement of mucosal folds and cobblestone appearance (Fig. 1). Those features most marked in the proximal bowel were thought to be consistent with extensive Crohn's disease although "lymphoma could not be ruled out".

To enable future investigations to be carried out it was felt that Mr McN required proper nourishment and total parenteral nutrition was commenced through the subclavian line. Three weeks later he developed superior vena caval obstruction by thrombus extending into the right atrium.

A hung perfusion scan showed evidence of multiple small pulmonary emboli. The thrombus was completely dissolved by treatment with BRC, a plasminogen tagged streptokinase.

Enteral feeding with an elemental diet soon produced diarrhoea. Crosby capsule biopsy revealed partial villous atrophy compatible with hypogammaglobulinaemia. After failure of a two week course of metronidazole to relieve his symptoms Mr McN eventually agreed to undergo laparotomy to provide a firm diagnosis. At laparotomy a two foot segment of small
bowel starting four inches beyond the duodenojejunal flexure was found to be thickened and narrowed (Fig.1a). Several skip lesions about three inches long were present over the next 18 inches. The ileum was grossly normal. As lymphoma could not be excluded macroscopically the first four feet of jejunum was resected and an end to end anastomosis performed. Pathological examination of the resected bowel reported a pattern of involvement and histological features suggestive of Crohn's disease but a number of characteristic features including granulomas, fissuring ulceration and transmural inflammation were absent. No neoplastic cells were visualised and immunohistochemistry demonstrated the presence of only T lymphocytes.

Following resection the patient became asymptomatic except for the occasional loose stool. He put on weight and serum total protein gradually returned to normal.

In April 1986 Mr McN presented with a 24 hour history of colicky abdominal pain and vomiting and had passed no stool since the onset. On examination he had mild central abdominal tenderness and foetid breath. Plain abdominal X-ray demonstrated some mildly dilated gas filled loops of bowel with gas present in the sigmoid colon. A diagnosis of subacute obstruction was made. Although he initially settled with fasting and antispasmodics his abdominal pain began to increase over the following three days and with no improvement in the abdominal films, it was decided to perform a laparotomy. A thick vascular band, causing obstruction in the mid-ileal region, was discovered. Two loops of ileum had undergone volvulus around this point, one showing early ischaemic changes. The band was divided and a side to side enterocenterostomy of the ileum proximal and distal to the ischaemic, compressed area, was performed. There was no evidence of recurrent small bowel disease. Apart from post operative wound and chest infections Mr McN made a good recovery and was discharged home.
Fig 1. Barium meal and follow through showing ulceration, tubular narrowing, sacculation and thickening of bowel wall together with effacement of mucosal folds and cobblestone appearance.
Fig. 1a. Resected specimen of jejunum.
Fig. 1b. Portion of resected specimen demonstrating ulceration and cobblestone appearance.
DISCUSSION

Patients with primary hypogammaglobulinaemia suffer from an increased incidence of malabsorption secondary to a variety of gastrointestinal complications including bacterial colonisation, giardial infestations and villous atrophy which may not be responsive to a gluten-free diet. Our patient presented with features of malabsorption associated with an ulcerative jejunitis observed on endoscopy. The most common cause of ulceration of the small intestine in Western society is Crohn's disease but the differential diagnosis includes tuberculosis, actinomycosis, typhoid, bacillary dysentery, polyarteritis nodosum, ischaemia, lymphoma and other neoplasms and the Zollinger-Ellison syndrome. With the majority of these conditions excluded a differential diagnosis of Crohn's disease and lymphoma remained.

A recent review of 96 patients with X-linked hypogammaglobulinaemia made no mention of the occurrence of Crohn's disease although a case of lymphoma affecting the terminal ileum was quoted (Laderman et al., 1985).

The clinical similarities between regional enteritis and lymphoma have long been recognised. Both may present with pyrexia, anorexia, weight loss, abdominal pain, malaise, anaemia and a palpable abdominal mass (Gray et al., 1982). In this clinical setting barium small bowel examination is often relied upon for clarification of the diagnosis. However on occasions these conditions are radiographically indistinguishable. This is not surprising since the human small intestine, as other organs, will respond macroscopically in only a limited number of ways to the multiple varied disease processes that may affect it. Thus on occasions different disease processes result in a similar gross pathological response. Both may produce strictures, demonstrate nodular patterns, aneurysmal dilatations, several types of ulceration, fistula formation, mesenteric masses and thickened mucosal folds (Sartoris et al., 1984).
The management of all patients with chronic inflammatory disorders of gastro-intestinal system involves close cooperation between the physician and the surgeon. With continuation of symptoms and no absolute exclusion of malignancy by radiological and histological means, exploratory laparotomy is indicated. At laparotomy the gross pathology in this case, being severely abnormal, could not be diagnosed with certainty although skip lesions, typical of Crohn's disease, were present (Figs. la, lb). The safest course of action to relieve symptoms and to remove a possible malignant lesion was followed.

Malignancy in the resected portion of bowel was excluded by special staining techniques of a full thickness biopsy, but the histology was also very uncharacteristic of Crohn's disease. In the sections examined there were no convincing granulomas or transmural fissures, the features seen in the early descriptions of the disease (Blackburn et al., 1939). However on closer inspection other features of Crohn's disease were seen, including, neuronal hyperplasia, focal hypertrophy of the lamina muscularis mucosae, pyloric metaplasia and the presence of inflammatory lesions in and around dilated lymphatics.

Another rare form of ulcerative enteritis associated with malabsorption, first described in 1949 (Nyman), must be considered, non-granulomatous ulcerative jejunoileitis (NGUJI). The condition presents as a chronic illness characterised by diarrhoea, steatorrhoea, weight loss and abdominal pain which is usually epigastric or periumbilical in site (Mills et al., 1980). Malabsorption is present in the majority of patients. The 'disease' has been reported in association with a variety of other conditions of the small bowel namely coeliac disease, non-responsive villous atrophy and lymphoma (malignant histiocytosis). Indeed some authors claim that most if not all the reported cases are examples of lymphoma (Isaacson
NGUJI has been described by Corlin and Pops in a single patient with primary hypogammaglobulinaemia whose symptoms responded to gammaglobulin therapy (Corlin and Pops., 1972), a course of action that gave no improvement in this case. Soltoft et al., (1972) have also reported a case of regional enteritis in a patient with selective IgA deficiency but as yet there have been no reports of case of NGUJI in patients with X-linked hypogammaglobulinaemia.

A definitive diagnosis remains to be made despite extensive discussion, but the occurrence of a Crohn's-like enteritis in a patient with complete agammaglobulinaemia raises interesting questions as to the pathogenesis of the inflammatory lesions in this case and in inflammatory bowel disease as a whole.

Both Crohn's disease and NGUJI are progressive with high incidences of recurrence after surgery and it was somewhat surprising, therefore, to note that no new areas of disease were found at laparotomy one year after the initial bowel resection.

The problems encountered in the surgical management of hypogammaglobulinaemic patients were well illustrated in this case. The inevitable problems of long term venous cannulation for total parenteral nutrition, especially a concern in a patient with malabsorption, and the high incidence of postoperative wound and respiratory infections, being the most obvious.
CASE TWO

Mr G.T., a 60 year old manager, was admitted on the Norfolk Hospital to Emergency by ambulance after an episode of severe gastrointestinal bleeding associated with the passage of large amounts of dark blood and a syncope attack. He gave a history of mild diarrhoea for approximately two months with the occasional passage of blood and mucus.

On examination the only finding of significance was a large effusion of the knee joint, now on the left side. Sigmoidoscopy revealed only mild inflammatory changes of the rectal mucosa but with some contact bleeding. Proctological examination of a biopsy of rectal mucosa was requested within seventeen hours. Radiological barium enema showed marked colonic abnormality that was reported by the radiologist to be due to changes of diverticulosis colored with irregularly arranged barium stained. The position was that some diverticulum was due to the nature of bowel motility. Treatment with an enema and prednisolone was continued with little improvement.

Mr G.T. was then seen in the Accident and Emergency Department of the Norfolk Hospital in emergency by ambulance and was pronounced dead. There were no symptoms of bowel obstruction, perforation, or ulceration. The post mortem showed a perforation in the sigmoid colon with a large abscess formation.

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CASE TWO

MR G.Y. (27.02.51)

Mr G.Y., a 35 year old brewery worker, first presented to the Princess Margaret Rose hospital in November 1972 with a painless effusion in the right knee associated with a persistent mild pyrexia and severe apthous ulceration of the mouth. On enquiry and examination there were no symptoms or signs of conjunctivitis, urethritis or diarrhoea. Erythrocyte sedimentation rate was 50 millimetres (mm) in the first hour. Serological testing for Anti-Nuclear factor was negative, and microscopy and culture of fluid aspirated from the joints showed no abnormalities. A urinary tract infection was discovered to be the cause of the pyrexia and was treated. No treatment was deemed necessary for the joint effusion, but a follow up appointment was made to review progress.

Mr G.Y. was next seen in the Accident and Emergency Department of the Royal Infirmary in February 1973 when he presented after an episode of explosive diarrhoea associated with the passage of large amounts of frank blood and a syncopal attack. He gave a history of mild diarrhoea for approximately two months with the occasional passage of blood and mucus. On examination the only finding of significance was a large effusion of the knee joint, now on the left side. Sigmoidoscopy revealed only mild inflammatory changes of the rectal mucosa but with some contact bleeding. Histological examination of a biopsy of rectal mucosa was reported within normal limits. Double contrast barium enema showed marked colonic abnormality from the hepatic flexure to the midpoint of the descending colon with irregular narrowing and marked mucosal oedema and ulceration; some ulcers appearing to penetrate into the bowel wall. Barium meal and follow through failed to display any signs of small bowel disease. Treatment with Salazopyrine and Prednisolone was initiated.
The patient's symptoms remained under control with this therapy, although the knee effusion remained and he developed a mild iron deficiency anaemia. On examination in May 1975 he was noted to have the rash of erythema nodosum on his right shin and mild haemorrhoids. A double contrast barium enema performed at this time displayed a typical picture of severe chronic ulcerative colitis involving the entire colon but sparing the sigmoid colon and rectum. A surgical opinion was sought with a view to colectomy but sigmoidoscopy to 25 centimetres (cm) revealed an essentially normal mucosa and Crohn's disease was thought a more likely diagnosis. It was decided to persevere with medical treatment.

Mr G.Y. remained relatively well on salazopyrine and prednisolone passing one or two loose stools per day. He developed a painful right hip which was diagnosed as sacro-ileitis which was relieved by a short term increase in the dosage of prednisolone. A double contrast barium enema performed in December 1977 showed continuing severe disease of the transverse and descending portions of the colon, but sigmoidoscopy again revealed no abnormality. Colectomy and ileorectal anastomosis was advised in view of the severity of the radiological changes, the systemic complications and the risk of malignant change. Mr G.Y. refused this option so treatment with salazopyrine alone was continued; it was felt to be unwise for him to continue on steroid therapy.

Salazopyrine proved to be sufficient to control the patient's symptoms. In January 1984, however, a full blood count showed Haemoglobin 10.8g/dl, White cell count $2.9 \times 10^9$/L, Platelets $97 \times 10^9$/L and Reticulocytes 7.4%; results consistent with a diagnosis of haemolytic anaemia, presumably with a side effect of long term salazopyrine therapy, which was therefore discontinued.
A double contrast barium enema was performed in September 1984 and displayed a long inflammatory stricture of the mid descending colon containing numerous pseudopolypi (Fig. 2). Colonoscopy confirmed the presence of the stricture containing widespread areas of ulceration and allowed biopsies of the numerous granuloma within a picture of active on chronic colitis. Interestingly a biopsy taken from macroscopically normal sigmoid mucosa revealed granuloma and giant cell formation.

Despite the severe changes in his colon, Mr G.Y. remained relatively symptom free, although his knee intermittently swelled and stiffened. During the months of May to August his diarrhoea returned along with a picture of iron deficiency anaemia (Haemoglobin recorded at 8.5g/dl at one consultation). This episode eventually settled with a treatment of steroids and Asacol.

In September 1985 Mr G.Y. developed a peri-anal abscess which was incised and drained. Discharge from the wound continued and an Examination Under Anaesthetic was performed in December. A fistulous tract was found passing through part of the lower sphincters to the anal canal. This was laid open and curetted free of all granulation tissue and packed with a Milton soaked wick. Histological examination of the curettings revealed the occasional granulomas and foreign body giant cells but no other features indicative of active Crohn's disease. Healing was slow to occur and the value of steroid and/or metronidazole therapy to aid this process was debated. A further exploration in April 1986 revealed a fistula extending to the level of the puborectalis muscle. The superficial part of the tract was laid open and a Seton placed around the remainder. Complete division of the tract would have been liable to result in incontinence. A referral to St Mark's Hospital, London, will probably be made.
Fig 2. Double contrast barium enema showing a long inflammatory stricture of the descending colon containing numerous pseudopolypi.
DISCUSSION

The extra-intestinal complications of inflammatory bowel disease are well known and include apthous stomatitis, uritis, skin lesions such as erythema nodosum and rarely pyoderma gangrenosum, cholangitis and seronegative inflammatory arthritis. Two patterns of arthritis are recognised; enteropathic synovitis and sacroiliitis sometimes progressing to ankylosing spondylitis. Enteropathic synovitis, the presenting feature in this case, is an acute, often migratory, non-erosive oligoarthritis occurring at some stage in 12% of patients with ulcerative colitis and 20% of those with Crohn's disease. The knees, ankles and other weight-bearing joints are most commonly affected but the wrists and small joints of the fingers and toes can also be involved. This arthritis however tends to follow exacerbations of the bowel disease, sometimes in association with apthous mouth ulceration and it is unusual for these manifestations to pre-empt the initial symptoms of inflammatory bowel disease. The second form of associated arthritis, sacroiliitis, also seen in our patient, occurs in 16% of patients. It pursues an independent course and often precedes the bowel disease. It is associated with a high prevalence of HLA-B27 while enteropathic synovitis is not. Our patient did not give a family history of HLA-B27 associated disorders.

It is only relatively recently that Crohn's disease of the large bowel has been recognised, with the first publication emphasising the pathological differences from ulcerative colitis appearing in 1960 (Lockhart-Mummery and Morson). Since then the clinical and radiological differences in the two disease entities have also been well described (Lennard-Jones et al., 1968; Marshak and Lindner, 1975). Diarrhoea is the most common presenting symptom, although not as common as in ulcerative colitis, often occurring with severe abdominal pain and the passage of blood per rectum. In 25% of patients the presenting symptoms are those of anal
lesions including fissures, fistulas, ulcers and abscesses; these complications occurring at some stage in their illness in 80% of patients (Lockhart-Mummery and Morson, 1964). The rectum is spared in 50% of cases, being an accepted feature of cumulative evidence for a diagnosis of Crohn's colitis (Schachter and Kirsner, 1975) and therefore sigmoidoscopy, as in this case, is often unrevealing. Double contrast barium enema examination will often assist the diagnosis. Seldom are there the signs of the uniformly contracted and shortened bowel with the loss of haustrations and granular mucosa characteristic of ulcerative colitis, it being more usual to see isolated changes. The bowel outline is more irregular with narrowing of the lumen and rigidity of the walls. In more severe disease, strictures, a 'cobblestone' mucosa, pseudopolyposis and deep fissures radiating outwards from the bowel lumen may be observed. The terminal ileum is involved in the disease process in approximately 30% of patients with Crohn's colitis and so must always be excluded by a barium meal and follow through examination.

Indications for surgery in Crohn's colitis are not as clear cut as those in the case of ulcerative colitis. Stenotic obstruction, toxic dilatation and perforation are absolute indications but opinions are divided as to the wisdom of surgery in cases of chronic ill health and acute episodes without dilatation. The results of a recent trial suggest that medical management may be preferable to surgery in acute Crohn's colitis (McIntyre et al., 1986). The effectiveness of a regimen of bowel rest, total parenteral nutrition and high dose steroid therapy was contrasted with one of an oral diet and high dose steroids. Although bowel rest had no effect on the outcome of a severe attack of ulcerative colitis with 50% requiring surgery, results tentatively suggested that this form of management may be all that is necessary to control Crohn's colitis. This relatively benign course of acute Crohn's disease has been confirmed
in other trials and adds weight to the advantage of distinguishing between ulcerative and Crohn's colitis as early as possible in the illness (Dickinson et al., 1980). However, the use of total parenteral nutrition remains very much under review. If the disease does cause severe disability with no remission of symptoms on medical management, or is complicated by severe systemic symptoms, then surgery may be considered. The principle of the surgical management of Crohn's disease, at any site, is conservation. If only a small segment of bowel is involved it may be reasonable to resect this locally with a margin of approximately 10 cm of normal bowel. The operation usually performed in diffuse colonic disease is a total colectomy with immediate or delayed ileorectal anastomosis. Goligher reports in a series of 48 patients in which a delayed ileorectal anastomosis was performed a 70% recurrence of disease in the ileum, 20% in the rectum and 10% in both (Goligher, 1979). These results were compared with those after treatment by permanent ileostomy because of minor changes present in the rectum. 70% of patients proceeded to suffer severe disease of the rectal stump of whom 35% required an abdominal-perineal resection, and only 15% had recurrence of the disease proximal to the stoma. Some authors now consider proctocolectomy and ileostomy to be the operation of choice in most cases (Heen et al., 1984), although major problems with this procedure are the frequent occurrence of a persistent sinus in the perineal wound, and troublesome skin excoriation around the ileostomy site (Spence and Wilson, 1983). All authors agree on the poor results of ileorectal anastomosis in Crohn's colitis with high incidences of anastomotic breakdown and recurrent disease, but since medical treatment may be effective in controlling recurrence or resection and reanastomosis possible it seems reasonable to many surgeons to offer restoration of gut continuation if there is a chance of success (Mortensen et al., 1984). This may be particularly important in early adult life when the presence of a stoma may be less acceptable than in later years.
One of the factors affecting the decision to operate in cases of ulcerative colitis is the potential for malignant change. Although early reports found no evidence for an increased risk of colonic cancer in Crohn's disease (Morgan, 1971), it is now recognized that this is not the case. An overall risk seven times greater than that of the general population has been reported (Greenstein et al., 1981) and a risk 20 times greater in those whose colitis began before the age of 21 years (Weedon et al., 1973). Most would not consider the overall risk significant enough to justify prophylactic surgery. However, these figures do justify close surveillance of the disease process to detect possible changes indicative of malignancy, a course of action that will be taken in the future management of the case presented here.

Anal lesions complicate the course of Crohn's colitis at some stage in approximately 60% of patients. Those most commonly occurring include fissures, oedematous tags and fistulae (Lockhart-Mummery, 1985). Some of those lesions heal with no active treatment especially if the intestinal disease is brought under control by medical or surgical means. Anal fistulae have been reported to occur in approximately 30% of patients with Crohn's colitis, the incidence rising as the site of the involved bowel becomes more distal (Hellers et al., 1980). 60% of anal fistulae in Crohn's disease are either superficial, simple intersphincteric or low trans-sphincteric, as compared to 75% of fistulae occurring in non-Crohn's patients. These may be managed by conventional techniques and although healing may be slower in Crohn's patients the results are very nearly as successful. Difficulties may arise when attempts are made to treat high, supra-sphincteric fistulae, similar to the one that has occurred in the patient presented. In these cases it is generally agreed that if continence is to be preserved surgical treatment should be limited to simple deroofing and decompression of the abscess cavity.
and curettage of the tracts. This may often be adequate to promote healing. There has been recent interest in the use of metronidazole and immunosuppression by steroids or azathioprine in the treatment of high fistulae, and this combination has produced definite, if slow, improvement in the more severe cases (Allan et al., 1977). Anal lesions are, however, a primary reason for the excision of the rectum in a third of cases where this proves necessary and in rare instances this may be due to the development of carcinoma within a chronic fistulous tract.
CASE THREE

My T.J. was admitted to the hospital on July 10, 1973, and underwent an operation to remove a benign polyp. The operation was performed under general anesthesia on July 12. The polyp was removed without complications.

On July 13, the patient was discharged from the hospital. He was advised to rest for one week and return for a follow-up appointment.

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he was found to be confused but apyrexial, to have hepatomegaly but no signs of chronic liver disease, a diffusely distended abdomen with shifting dullness and to be tender across the lower abdomen. Bowel sounds were present. Stools were positive for occult blood. On investigation a plain abdominal X-ray showed a distended colon with mucosal degeneration and a provisional diagnosis of toxic megacolon was made. Liver function tests were still abnormal and given that there were no signs of chronic liver disease a diagnosis of sclerosing cholangitis was made. A treatment regime of four litres of fluid per 24 hours, hydrocortisone 100 milligrammes six hourly and antibiotic therapy with cefotaxime and metronidazole was instituted. A surgical opinion was sought. An abdominal X-ray on the following day showed the transverse colon to be dilated further to eight centimetres (see Fig. 3). He was passing about six fluid stools per day at this stage and it was decided that medical management should be continued to correct the hyponatraemia (125mmol/L) and the relatively low protein (52g/L). He became less confused although being incontinent of several fluid stools per day and being still clinically dehydrated. He was transferred to a surgical ward on the 22nd April in case of rapid deterioration but no indication for surgery was felt to be present at this stage. A central feeding line was inserted and total parenteral feeding at half strength commenced.

On the 23rd April Mr T.J. showed some signs of improvement; on X-ray the colonic dilatation was less marked and on examination his abdomen was non tender and bowel sounds had returned having been absent for four days. However, on the 24th April he began to have increasing diarrhoea associated with the passage of frank blood, he complained of feeling very tired and on examination he was obviously unwell. On examination the only positive finding was a mild tachycardia of 100 beats per minute. Blood analysis revealed a prothrombin ratio of 2.5. A coagulation screen
was then performed with the following results; PTR 5.5, PTTK patient 97 seconds, control 29 seconds, FDP's 80-160mg/L, Platelets 72x10^9/L and fibrinogen 0.34g/L. A diagnosis of disseminated intravascular coagulation (DIC) was made. Other abnormal results included bilirubin 122 umol/L, protein 43g/L and urea 20.8 mmol/L. Deterioration was thought to be due to fulminant colitis although apparently improving and no X-ray signs of perforation and by superinfection, for example by Clostridia organisms leading to DIC. Samples of blood and stool was sent for microscopy and culture. The DIC was treated with two units of fresh frozen plasma, six units of cryoprecipitate, six units of platelets and one unit of red cell concentrate to correct the anaemia. Antibiotic therapy with gentamicin, aslocillin and an increased dose of metronidazole was instituted. It was felt at this stage that colectomy was now indicated although he was still metabolically unstable, total protein was now down to less than 19g/L. However, before surgery could be performed Mr T.J. became progressively unresponsive, his respiratory rate and pulse rate decreased, his blood pressure became unrecordable and death rapidly followed.

Post mortem examination revealed a grossly abnormal colon with large areas of ulceration and pseudopolyposis. Active bleeding was noted from a large duodenal ulcer and from acute oesophagitis. The liver demonstrated changes characteristic of sclerosing cholangitis. Microscopy and culture of the specimens taken on the day that death occurred could not demonstrate any possible pathogen.
Fig 3. Plain abdominal X-ray showing dilatation of the transverse colon to 8cm with extensive mucosal degeneration.
DISCUSSION

The association of ulcerative colitis and liver disease was first described by Thomas in 1874 and later confirmed by Lister (1899). It has been estimated that in approximately 8% of patients with ulcerative colitis serial biochemical abnormalities, indicative of liver disease, occur at some stage of their illness (Dew et al., 1979). Pericholangitis is the most commonly encountered disorder with sclerosing cholangitis, cirrhosis and biliary tract carcinoma being important but rare.

Sclerosing cholangitis consists of fibrotic obliteration of the large bile duct system, often in association with intralobular changes, focal necrosis, Kupffer cell hyperplasia and perilobular fibrosis. The medium age of onset of hepatobiliary disease in patients with ulcerative colitis has been shown to be approximately 25 years (Schumpf et al., 1982). In the majority of patients the first signs of the development of sclerosing cholangitis, as occurred in the patient presented here, are an increase in the levels of alkaline phosphatase, alanine aminotransferase and aspartate aminotransferase with the bilirubin level elevated only in a minority. All patients exhibit changes on endoscopic retrograde cholangiography and indeed this is the most sensitive method of monitoring the progression of the disease. Multiple short stenoses of both intra- and extrahepatic bile ducts are characteristically seen. Unfortunately colectomy does not bring about remission of the disorder as is the case in many of the systemic complications of ulcerative colitis.

Interestingly the tissue antigens HLA B8 and HLA DR3 are found in 80% and 70% of patients, respectively, with combined ulcerative colitis and hepatobiliary disease. This finding may be relevant to the patient's strong family history of inflammatory bowel disease.
It was not possible to discover any reports of toxic dilatation of the colon occurring as a complication of an acute exacerbation of sclerosing cholangitis. Toxic dilatation has been defined by the radiographic demonstration of dilatation of the transverse colon to greater than 8 cm in diameter in a patient who is severely ill with colitis (Schofield, 1982). Others have used a diameter of 6 cm associated with the radiological features of bowel wall thickening, mucosal oedema and thumbprinting and, frequently, a paucity of gas in the distal colon (Spence and Wilson, 1983).

The exact pathogenesis of the condition is unknown, one early report suggested that transmural oedema interferes with the intrinsic mucosal plexus of the colon and thus compromises motility (Lumb et al., 1955). Superimposed infection, particularly by Salmonella species, and the use of antispasmodic agents have also been implicated in the development of dilatation (Schofield et al., 1979). It is therefore always important to exclude a relatively easily treated infective cause of toxic dilatation.

Clinical features of toxic dilatation include abdominal distension with diffuse tenderness, frank blood in the stools, abnormal appearances at sigmoidoscopy and signs of systemic toxicity. Various parameters have been used to assess the severity of acute colitis. Truelove and Witts (1955) used bowel frequency, temperature, pulse rate, haemoglobin concentration and the erythrocyte sedimentation rate and designated attacks as mild, moderate and severe. Lennard Jones et al. (1975), however, found that a combination of bowel frequency and body temperature in the first 24 hours after admission provided the best predictive value as to whether a patient would respond to medical therapy or not. Conservative management with intravenous fluids and steroids may be effective in up to 50% of patients with acute colitis (McIntyre et al., 1986) but many authors regard toxic dilatation as an absolute indication for surgery. Ritchie et al., (1984) reported a mortality of 59% in patients in whom toxic dilatation had been present for two days or more before surgery...
and a plea was made for prompt operative treatment if this complication arose. Emergency surgery for ulcerative colitis does however itself carry a high mortality rate, most series quoting a figure of approximately 20% (Koudahl and Kristensen, 1976) and thus the importance of resuscitation producing biochemical and haemodynamic stability is clear. Unfortunately adequate stabilisation proved impossible in this case with co-existent hepatobiliary complications and the onset of disseminated intravascular coagulation.

Disseminated intravascular coagulation (DIC) has been defined as a "clinico-pathological syndrome of variable expression, resulting from uncontrolled simultaneous activation of the coagulation and fibrinolytic systems" (Bell, 1980). It has been the subject of extensive literature in past years and although much is known of the pathogenesis of the disorder many of the mechanisms resulting in the clinical and laboratory features of this illness are unknown. The most probable etiologic factor responsible for inducing DIC is the entrance of a thromboplastic substance into the circulation (Hardaway et al., 1976). This may be produced by areas of damaged, hypoxic or infarcted necrotic tissue as is present in acute ulcerative colitis. Endotoxin and other toxins released from microorganisms are also capable of inducing DIC (Berghaus and Lohmann, 1974). Superinfection of the acute colitis presented here possibly being the causative factor. DIC is also frequently observed in association with liver disease of all recognised categories. Ulcerative colitis is rarely mentioned as a primary cause of DIC (Minna et al., 1974) but any severe illness may induce this condition and several possible causative factors were present in the patient presented. Although the clinical features are usually quite striking, being most commonly a tendency to bleed or actual bleeding from multiple sites including the skin, mucous membranes, trachea, gastrointestinal tract and genitourinary tract, the diagnosis
of DIC is made by laboratory investigation. It is necessary to show
depletion of coagulation factors and platelets and evidence of an increase
in the products of fibrinolytic activity (fibrin degradation products).
In general the most important principle in the management of patients
with DIC is prompt identification reversal and eradication of the under-
lying illness. If this cannot be accomplished the chances for successful
elimination of DIC and its consequences become severely limited. Along
with measures to maintain circulatory volume, organ perfusion pressure,
electrolyte/acid base balance and tissue oxygenation, platelets, fresh-
frozen plasma and cryoprecipitate (an excellent source of fibrinogen)
can be given to replace coagulation factors. Death may be caused by
unexpected exanguination into a body cavity or more frequently due to
pulmonary failure, septicaemia, cardiogenic shock, acute tubular necrosis
and hepatic failure. In the vast majority of patients, however, autopsy
data suggest that death was due to an unremitting, underlying disease
process.
CASE FOUR

A troponin I level was measured on admission, which was 0.01 ng/mL. The patient had a high-sensitivity troponin level of 2.3 ng/mL. A chest X-ray showed no cardiac enlargement, and an ECG was normal. An echocardiogram showed normal left ventricular function.

During hospitalization, the patient developed fever and chills. A white blood cell count was 12,000 cells/mm³, and the patient was started on broad-spectrum antibiotics. A CT scan of the abdomen showed an abscess in the right lower quadrant. The patient underwent an exploratory laparotomy, which revealed a perforated appendix. The appendix was removed, and the patient was treated with antibiotics and intravenous fluids.

The patient made a complete recovery and was discharged home on the 10th hospital day. The wound was closed with a surgical stapler and a drain was left in place for 24 hours. The patient was seen in the clinic 2 weeks later, and the drain had been removed. The wound was healing well, and the patient had no fever or abdominal pain. The patient was advised to continue taking antibiotics for 10 days and to return for a follow-up visit in 1 week.
CASE FOUR
MIS S.C. (19.12.63)

Miss S.C., a 22 year old nurse, first presented to the physicians in April 1985 with a four month history of increasing diarrhoea, occasionally occurring up to 12 times per day. The diarrhoea was associated with cramping lower abdominal pains and the passage of blood and mucus. On enquiry into her past history she was noted to have had a normal appendix removed in spring 1985 with no other causative lesion discovered for an episode of lower abdominal pain. She claimed a long history of recurrent conjunctivitis and aphthous ulceration of the mouth. More recently associated with the diarrhoea she reported nausea and occasional vomiting, amenorrhoea, a weight loss of one and a half stone and an itchy rash covering the extensor surfaces of her elbows, shoulders and back. She is also asthmatic, controlled on treatment with salbutamol and beclamethasone. Examination findings were unremarkable other than marked tenderness on rectal examination. Sigmoidoscopy displayed an oedematous and friable rectal mucosa producing large quantities of mucus but with no evidence of ulceration. A biopsy of rectal mucosa was reported a showing changes strongly suggestive of Crohn's disease. On blood analysis, significant results were an erythrocyte sedimentation rate of 32 millimetres on the first hour and a haemoglobin level of 10.9g/dl. A double contrast barium enema revealed a colitis extending from the caecum to distal sigmoid colon with sparing of the rectum (Fig. 4). Ileocaecal reflux was noted, the terminal ileum being abnormally dilated, although there was no evidence of a mass, as is often seen in cases of Crohn's disease.

Although being initially controlled on salazopyrine alone, further recurrences in July and August 1985 required the addition of prednisolone to the regimen. A barium meal and follow through examination in August failed to demonstrate any small bowel involvement.
After further exacerbations in August and September, in which she presented at the Accident and Emergency department, a surgical opinion was sought. It was noted that although her symptoms seemed severe, faecal occult blood testing had proved negative and all biochemical parameters including protein level were within the normal range despite the apparent severity of her diarrhoea.

A double contrast barium enema carried out later in September revealed generalised distension of the colon with a granular and ulcerated mucosa of the descending portion. Miss S.C. was admitted for assessment in October 1985. Colonoscopy was performed and the typical features of ulcerative colitis with mild to moderate inflammation were noted, involving the entire colon. Biopsies were taken from the caecum, hepatic flexure, transverse colon, descending colon and sigmoid colon. Histological examination was reported as showing a picture of acute on chronic ulcerative colitis. Faecal occult blood testing again proved negative which is unusual in cases of diarrhoea due to inflammatory bowel disease. Surgical intervention was deferred.

However, during her admission, she became pyrexial with accompanying tachycardia, increasing lower abdominal pain, diminishing bowel sounds and a plain abdominal X-ray demonstrated a transverse colon dilated to a diameter of six centimetres. Treatment with intravenous steroids was instituted but her condition rapidly deteriorated over the following few hours and emergency total colectomy and ileostomy was performed. The rectum was simply closed, a pre-operative sigmoidoscopy showing virtually normal rectal mucosa. Macroscopically the resected bowel appeared markedly dilated throughout with the appearances of active ulcerative colitis. Histologically, however, the appearances were those of Crohn's disease, granulomata and submucosal extension of disease noted. Although
as a whole the features were thought to be not entirely typical and may have been modified by steroid treatment.

The patient's immediate post-operative recovery was good; her ileostomy began to function well and by day four she was eating. However, on day five she became distressed, complaining of nausea, retching and gaining no relief from her abdominal pain by the prescribed analgesia. Her stoma was reported as appearing dusky. Her white blood cell count over a period of three hours rose from 7.2 to $34 \times 10^9/\text{L}$. She was returned to theatre. At laparotomy free serous fluid was present in the peritoneum and from a point about 45cm distal to the duodenojejunal junction the whole small bowel displayed marked ischaemic changes, being plum coloured, oedematous and failing to exhibit peristalsis on stimulation. The proximal 45cm showed a patchy blue discolouration but was generally pink. There was no evidence of major venous thrombosis, internal herniation or volvulus. Poor pulsation of the superior mesenteric artery was noted which improved after a fruitless exploration, proximally and distally with a Fogarty catheter. Warm packs improved the condition of the proximal bowel but the distal portion was clearly non-viable. Serial incisions were made in the bowel wall working proximally until free bleeding of oxygenated blood occurred. The bowel was transected at this point, (approximately the midpoint of the small bowel) and the cut end brought out through the previous ileostomy site and a "second look" laparotomy arranged for the following day. The patient's condition deteriorated and at laparotomy further necrosis had occurred although pulsation in the mesenteric vessels appeared very satisfactory. Further bowel was resected and a jejunostomy was fashioned at a point 30cm from the duodenojejunal flexure. No explanation could be found for the sequence of events and histological examination of the resected bowel was unrevealing, there being no evidence of a vasculitis of inflammatory bowel disease.
Miss S.C. made a good post-operative recovery and a subclavian feeding line was inserted. Over the ensuing weeks, however, the line had to be removed and resited on three occasions because of infective problems. Since February 1986 Miss S.C. has been discharged home for three to four days at a time and is then admitted for two days for a course of intravenous fluids to correct the metabolic upsets which inevitably occur. Haemoglobin levels are well maintained with vitamin B12 replacement therapy and serum protein levels remain stable. Her main problems are with sodium and magnesium depletion, despite oral replacement, which have caused disturbing neuromuscular symptoms. Advice concerning her future care, especially as regards the possibility of home parenteral nutrition, has been sought from the Nutrition Unit at Salford.

Disturbingly, a rectal biopsy performed in April 1986 showed evidence of active Crohn's disease.
Fig 4. Double contrast barium enema showing a colitis extending from the caecum to the distal sigmoid colon. There is a shallow ulceration, a smooth poorly coated mucosa, a degree of ileocaecal reflux and generalised loss of haustrations.
In most series of patients with non-specific inflammatory large bowel disease a confident diagnosis cannot be made in 10-15% of cases (Glotzer et al., 1970; Price, 1978). Although a differential diagnosis can usually be made on clinical, pathological and radiological evidence (Lennard-Jones et al., 1968), in many cases difficulties arise due to overlapping in the pathological features of Crohn's disease and ulcerative colitis. Indeed it has been suggested that these two syndromes may be opposite ends of a spectrum of one disease (Lewin and Swales, 1966). Although pathological features such as fissuring ulceration, transmural inflammation and a maintained goblet-cell population may occasionally be present in ulcerative colitis the granulomas seen on histological examination of the resected bowel in this case are considered to be pathognomic of Crohn's disease. The rectal sparing seen here can also be taken as evidence for a diagnosis of Crohn's disease (Schachter and Kirsner, 1975). Macroscopically, in acute disease, appearances are often very similar (Price, 1978).

Toxic dilatation of the colon is a phenomenon which was initially described as complicating ulcerative colitis, but as Crohn's disease of the colon became increasingly recognized it was realised that it could also be a complication of this disease (Turnbull et al., 1970). In a recent series this complication was seen to arise in approximately 10% of patients (Spence and Wilson, 1983). Although bowel rest in conjunction with high dose intravenous steroids is often sufficient to alleviate symptoms in acute Crohn's colitis (McIntyre et al., 1986), the refractory nature of the episode described in the patient presented, and the development of toxic dilatation necessitated surgical intervention. Surgery in the treatment of toxic dilatation is the same whatever the underlying diagnosis (Mortensen et al., 1984). Although Turnbull et al. (1971) have advocated treating severe toxic megacolon by ileostomy coupled with single
or multiple "blow-hole" colostomies, the operations of choice at present are either colectomy or panproctocolectomy accompanied by an ileostomy. Poor results from immediate ileorectal anastomosis following excision for toxic dilatation have been described in both ulcerative colitis and Crohn's disease (Buzzard et al., 1974). The choice between colectomy and panproctocolectomy has been hotly debated. Proctocolectomy was advocated by Lee and Truelove (1980) who confirmed earlier reports that this operation does not carry a greater mortality than simple colectomy and indeed some series showed a lower mortality (Scott et al., 1970; Jones et al., 1977). The operation of proctocolectomy may be more attractive in cases of toxic dilatation complicating ulcerative colitis, there being and large incidence of late complications involving the rectal stump, not least being the development of malignancy, requiring excision (Moss and Keddie, 1965). However, in this case, there being no obvious involvement of the rectum, and considering the age and sex of the patient, colectomy and ileostomy with the hope of a future anastomosis would seem the operation of choice. Although recurrence is likely in the rectum (Goligher, 1979), this will in all likelihood respond to medical treatment, but an in situ rectum may always be a latent reservoir for the late complications of spontaneous perforation and carcinoma (Weedon et al., 1973).

Emergency colectomy for acute colitis is associated with many post operative complications, however ischaemia of the bowel has not been listed among them. The most common cause of acute ischaemia is thrombotic or embolic occlusion of the superior mesenteric artery. In most cases this occurs in patients with diffuseatheroma or with a recognisable source of emboli such as intracardiac thrombus forming in atrial fibrillation or after a recent myocardial infarction. Venous occlusion is a less common cause and may occur in pregnancy, the puerperium, in young women taking oral contraceptives (our patient was not using these) and in those predisposed
to intravascular coagulation. No evidence of either of these conditions was found at laparotomy in this case. Non-occlusive mesenteric ischaemia (NO MI) first described in 1958 (Ende, 1958) has recently been reported to be responsible for 25-50% of all mesenteric vascular accidents (Brandt and Boley, 1981). The pathogenesis is believed to involve splanchnic vasoconstriction occurring in response to a decrease in cardiac output, hypovolaemia, dehydration, vasopressor agents or hypotension. This vasoconstriction may persist even after the initiating cause has been corrected. Predisposing conditions include myocardial infarction, congestive cardiac failure, renal and hepatic disease and most relevantly major abdominal or cardiac operations. The condition generally occurs in the elderly in whom it carries at 90% mortality. It may be diagnosed angiographically by signs of mesenteric vasoconstriction and if this investigation is performed early enough a papaverine infusion may be curative. Other diagnostic features are common to other ischaemic syndromes, including, a leukocytosis often greater than 15 000 cells per cubit millimetre, metabolic acidosis with an increasing base deficit, pain is a relatively uncommon feature especially in NOMI and unexplained abdominal distension or gastrointestinal bleeding may be the only clinical finding. The decreased mesenteric blood flow of NOMI is not amenable to surgical correction and indeed manipulation of the superior mesenteric artery at laparotomy should be kept to a minimum. Resection of ischaemic bowel and primary anastomosis with a second look operation 24 hours later is the recognised form of management. The extent of the resection necessary in the case reported presents many more problems of management.

Most adults have approximately six metres of small intestine. As a general rule when three metres or less remain a number of serious metabolic and nutritional abnormalities occur, with less than two metres most patients have a limited work capacity as well and many with less than one metre
of small bowel remaining require parenteral nutrition at home on an indefinite basis (Hill, 1985). When patients are left with a jejunostomy the remaining bowel cannot concentrate luminal contents so that isotonic water and salt loss is severe resulting in dehydration, sodium depletion, hypocalcaemia and hypomagnesaemia. Zinc deficiency may also become a problem. Removal of the ileum results in a decreased absorption of bile salts and vitamin $B_{12}$, leading to a decreased absorption of fats and fat-soluble vitamins and pernicious anaemia. Although the remaining bowel has a remarkable ability to adapt these specific absorptive functions are never regained. This adaptive process continues for one to two years and involves gross mucosal hypertrophy with an associated increase in absorptive function (Bristol and Williamson, 1985). This process is probably overwhelmed if there is less than one metre of small bowel remaining. Interestingly it is also associated with an increase in carcinogenesis so patients must be followed up with this potential complication in mind. Many other less drastic but more common complications of massive enterectomy have been reported. Troublesome excoriation around the stoma site may accompany the large fluid loss often seen in the immediate post-operative stages. Electrolyte abnormalities such as the hypomagnesaemia encountered in our case may present acutely with severe neuromuscular symptoms and associated sodium, water and calcium depletion (Selby et al., 1984). Long term complications include an increased risk of urinary tract calculi of all types associated with a low urine volume, decreased calcium excretion and increased concentration of urinary oxalate (Bambach et al., 1981). Osteomalacia is said to occur in approximately 30% of patients after extensive enterectomy (Compston et al., 1978).

Although the patient described has shown evidence of intestinal adaption she still poses severe problems in the stabilisation of electrolyte balance. The available options for management include home parenteral
and home enteral nutrition. A series of patients managed with total parenteral nutrition at home for at least a while showed few complications and a good work and social rehabilitation (Jeejeebhoy et al., 1973). Recently nocturnal nasogastric tube feeding to supplement a daily oral diet has achieved good results in patients with very little small bowel remaining (McIntyre et al., 1983). This method of treatment may warrant a trial in this case. Recent advice has suggested that since this patient's main problem is electrolyte balance she might benefit from the Portacath system. This consists of a completely implantable cannulation system which is easily accessed by an ordinary syringe needle connected to an infusion set. Patients are encouraged to infuse themselves with saline as rapidly as possible to minimise the inconvenience of an infusion.

Finally, small bowel transplantation may offer a future alternative to patients with short bowel syndrome. However, the large amount of lymphoid tissue within the small bowel is a potent antigenic stimulus for the rejection response and currently available immunosuppressive regimens are not adequate to prevent this arising (Pritchard and Kirkman, 1985).
CASE FIVE

Mr All, a retired medical officer, was admitted to hospital in December 1974 with a complaint of diarrhea. Double contrast barium enema showed a colitis from the rectum through to the terminal ileum. He was treated with a mild purgative, but the diarrhea persisted, and a medical opinion was obtained as to the possibility of a neoplastic lesion. A barium enema was repeated on an outpatient basis.

Mr All was admitted again in January 1975. The diarrhea was still present, and a medical opinion was obtained as to the possibility of a neoplastic lesion. A barium enema was repeated on an outpatient basis.

The patient was discharged and followed up on an outpatient basis.
Mr A.H., a schoolteacher, presented to the physicians in 1974 with a short history of diarrhoea accompanied by the passage of blood per rectum. After sigmoidoscopy and barium enema a diagnosis of localised proctitis was made and treatment comprising salazopyrine and predsol enemas was initiated. He had one further stress related exacerbation of his proctitis in December 1974 which settled with steroid treatment.

His proctitis remained under control with salazopyrine until 1978 when he was admitted with a severe, acute, onset of abdominal pain and bloody diarrhoea. Double contrast barium enema showed features of ulcerative colitis from the mid transverse colon distally with marked pseudopolyp formation. He was treated with high dose steroids for several weeks and a milk-free and then an elemental diet was introduced to attempt to gain relief of symptoms. He settled eventually after a two month stay in hospital and was discharged on salazopyrine therapy.

Mr A.H. was admitted again in August 1980 with similar symptoms. The diarrhoea was quickly brought under control by steroid treatment, but symptoms recurred as soon as the dose was dropped below 15 milligrammes (mg) per day. Relapses also accompanied the slightest stress or upper respiratory tract infection.

The patient's disease was investigated fully in the early months of 1981. Double contrast barium enema revealed severe disease with pseudopolyp formation in the descending and sigmoid colon. Sigmoidoscopy displayed a granular mucosa with ulceration, pseudopolyposis, contact bleeding and increased quantities of mucus. Biochemistry and haematology showed a mild degree of hypoproteinaemia and an anaemia of iron deficiency type.
Barium meal and follow through revealed no signs of small intestinal involvement. No pathogens were isolated from the faeces. Mr A.H. felt the symptoms of his colitis were interfering with his work as a teacher and the time involved in treatment adversely affecting his chances of promotion. It was felt that a surgical operation was necessary.

Mr A.H., however, was admitted from the Accident and Emergency department in May 1981 before his appointment with the surgeons was due. He complained of the passage of loose stools associated with blood and mucus approximately 10 times per day for the previous few days, accompanied by spasmodic abdominal pain which required pethidine for adequate analgesia. Sigmoidoscopy revealed a rectal mucosa that was not as abnormal as expected. This finding along with an apparent history of perianal infections and a lack of a histological diagnosis of ulcerative colitis raised some suspicion of the possibility of Crohn's colitis. Symptoms were of such a degree and of such a long standing nature that surgery was felt to be warranted. A colectomy was performed, macroscopic examination of the rectal mucosa revealed a cobble-stone appearance typical of Crohn's disease and the rectum was brought out at the lower end of the incision as a mucus fistula. An ileostomy was fashioned. Histology of a rectal biopsy reported active, severe ulcerative colitis with no granuloma formation.

Post-operative recovery was excellent but he continued to produce a fair quantity of mucus from his fistula.

In December 1982 he presented with symptoms of perianal abscess and on operation for incision and drainage a large fistulous track between the rectum and the abscess was discovered. A recurrence of his proctitis was also noted at operation and a biopsy of rectal mucosa demonstrated
active on chronic proctitis and mild epithelial dysplasia. Salazopyrine therapy was continued and follow up scheduled for six month intervals.

Biopsies were taken at each consultation and no further dysplasia was noted. In September 1985 sigmoidoscopy revealed an oedematous mucosa with contact bleeding and in view of the time lapse from the first diagnosis, the possibility of removal of the rectum was discussed with Mr A.H. A loopogram (Fig. 5) confirmed the presence of active colitis within the rectum, demonstrating a granular mucosa and several small polyps.

Sigmoidoscopy in April 1986 revealed a fibrotic and bleeding rectal stump and on consent from the patient excision is planned for September 1986.
Fig 5. Double contrast barium loopogram showing active colitis within the rectum, a granular mucosa and several small polyps.
DISCUSSION

The case reported here again demonstrates some of the difficulties involved in differentiating between ulcerative colitis and Crohn's disease of the colon. During the chronic phase of the disease reliable diagnostic features will often be found on histological examination of biopsy material. Unfortunately biopsies were not performed as part of the diagnostic process in this case; a diagnosis of ulcerative colitis being made on clinical and radiological grounds. In the acute phase of the disease, however, the pathology of Crohn's colitis and ulcerative colitis have much in common (Price, 1978). This finding has led Price to apply the term 'colitis' to the small group of cases in which there was difficulty in making an absolute diagnosis from the excised specimen. This group consisted of 10-20% of cases of non-specific inflammatory bowel disease. Rectal sparing and a cobblestone appearance of the mucosa, seen in this case for example, although features of Crohn's disease in the chronic state, have been observed in some indeterminate cases in the acute stage, subsequently proved to be ulcerative colitis. On the other hand, although the pathognomonic feature of Crohn's disease, the presence of granulomas, occurs in 60% of cases in the chronic state, granulomas are only found in 25% of cases on acute presentation (Glass and Baker, 1976). In this case, however, it is probable that a diagnosis of true ulcerative colitis can be made. A diagnosis which may be had some bearing on surgical management.

There are two main reasons for the removal of the colon in chronic ulcerative colitis. First, if the patient is much troubled by repeated attacks of the illness or if the symptoms are continuous in spite of good medical therapy, it is often advisable to remove the colon. Secondly, colectomy is sometimes necessary to deal with the complications or possible complications (malignancy being the most obvious) of chronic disease (Lee
The features of this patient's colitis and the possible long term complications of the high dose steroid therapy necessary for its control, fulfilled these criteria amply. The admission of the patient with an acute relapse was the deciding factor as to the time scale of operative intervention.

The choice of operation for chronic disease, as for toxic dilatation, is the subject of some debate. The main contenders being colectomy with immediate or delayed ileo-rectal anastomosis, and proctocolectomy with ileostomy. Opinion is divided, both operations having advantages and disadvantages. Ileorectal anastomosis is attractive to most patients since life with an ileostomy is avoided. Aylett, the main proponent of ileorectal anastomosis, employs a primary anastomosis however inflamed the rectal mucosa may be. Most surgeons will perform an ileostomy and leave the rectal stump as a mucous fistula to give the rectal mucosa chance to improve before the anastomosis is made. Aylett stated that up to 94% of patients are satisfied with the results (Aylett, 1963), but later he found a disturbing incidence of carcinoma in the retained rectum (Aylett, 1971). This has been confirmed by others (for example, Moss and Keddie, 1965). These rectal tumours have a very poor prognosis. Some surgeons advocate ileorectal anastomosis particularly in young patients, but it must be remembered that ulcerative colitis beginning at a young age carries a particularly high risk of cancer. Other disadvantages of retention of the rectum, with or without anastomosis, are that the inflammation usually persists and may become worse, with severe symptoms and the risk of local and remote complications of ulcerative colitis such as abscesses, fistulas, arthritis, uveitis and skin lesions. In one series 72% of patients required removal of the rectal stump after a follow up period of 25 years from the time of colectomy (Moss and Keddie, 1965). The operation of proctocolectomy and permanent ileostomy, because
of the described complications is becoming the operation of choice. It has no greater a mortality than the less extensive operations and removes the whole source of potential complications. Proctocolectomy is however a radical operation and a procedure such as rectocolectomy with anal conservation may become more popular (Deane and Celestin, 1983). This technique avoids the problems associated with perineal resection such as delayed healing, difficulties in micturition and sexual dysfunction. It is of utmost importance, however, to be aware of the social, psychological, sexual and practical difficulties faced by patients after ileostomy. This is of importance both in deciding which form of operation is to be preferred in a particular patient, and in counselling those who are to be given a permanent ileostomy.

The increased risk of developing carcinoma in a retained rectum after colectomy has been mentioned briefly. The cumulative frequency has been estimated at 6% at 20 years and 15% at 30 years (Grundfest et al., 1981). Although this is much less than the overall risk of cancer in ulcerative colitis, surveillance of the state of the rectal mucosa is mandatory. Mild epithelial dysplasia, as seen in this case, often occurs from time to time in ulcerative colitis. Had this progressed to intra-epithelial neoplasia during close surveillance, this would have been an indication for removal of the rectum (Riddell, 1985).

Unfortunately, symptomatic disease of the retained rectum has continued in this patient and excision of the rectal stump, to alleviate symptoms and to prevent further complications, is the obvious course to be taken.
CASE SIX

Dr. A.C. was referred for a marginal upper GI series. The patient presented with a history of a 2 month upper GI bleed. He had been on H2 blockers for several months and was also taking iron supplements. On physical examination, he appeared pale and anemic. A blood transfusion was ordered, but the patient's condition did not improve. The marginal upper GI series revealed a vascular malformation in the stomach. The patient was referred to a gastroenterologist for further evaluation.

The patient's symptoms were well controlled with iron supplementation and regular blood transfusions. However, exacerbations occurred with stress, and he continued to take H2 blockers daily. He kept regular follow-up appointments and was monitored closely. The patient eventually underwent a gastric resection, and his symptoms improved significantly.

In conclusion, the patient's case highlights the importance of early intervention and regular follow-up in managing marginal upper GI bleeds. The use of iron supplements and blood transfusions was crucial in managing the anemia, but further investigation into the vascular malformation was necessary to prevent future bleeds.
CASE SIX

MR A.C. (05.10.44)

Mr A.C., a 41 year old town planner, first presented to the physicians in 1970 with a short history of severe diarrhoea 10-12 times per 24 hours associated with the passage of mucous and blood. This episode coincided with the attainment of a new job and the development of mental illness in his mother with whom he lived. Double contrast barium enema and sigmoidoscopy revealed no abnormality and his symptoms were treated successfully with anti-diarrhoeal agents.

Mr A.C. was referred for a surgical opinion after a more severe recurrence of symptoms one year later. Sigmoidoscopy revealed a severe haemorrhagic proctocolitis. Mucosal biopsy showed superficial ulceration, acute on chronic inflammation and crypt abscesses on histological examination; features of non-specific proctitis. This acute episode was successfully managed by treatment with salazopyrine, predsol retention enemas and prednisolone suppositories, and a maintenance course of salazopyrine continued.

The patient's symptoms were well controlled on salazopyrine therapy, exacerbations occurring with trials of cessation of treatment and at times of stress at his place of work and in the care of his mother. He was kept under regular review, rectal biopsies and double contrast barium enemas being performed every two years. By 1978 double contrast barium enema exhibited changes in whole colon distal to the hepatic flexure, there being loss of haustrations associated with a granular mucosa.

In 1982 an exacerbation, associated with the failing health of his mother, required steroid treatment for its control although changes in the rectal mucosa visualised by sigmoidoscopy were slight in comparison to his
symptoms. After this episode it was necessary to continue low dose steroid therapy to control symptoms.

An in-patient assessment of the state of Mr A.C.'s colitis in August 1985 revealed little. Changes viewed on double contrast barium enema, although the bowel was poorly prepared, seemed to indicate little advancement of the disease since 1978 and sigmoidoscopy displayed an essentially normal rectal mucosa. However, at this stage Mr A.C. noted that symptoms were only controlled by a dose of at least 15mg of prednisolone a day.

After a General Practitioner referral to the Western General Hospital Mr A.C. was placed on a trial of a dairy product free diet in January 1986. This produced excessive flatulence, three bulky motions per day and a fissure-in-ano!

On review in February it was noted that the patient was acutely uncomfortable on rectal examination, with considerable spasm of the anal sphincter. There was a suspicion of a mass felt but it was thought to be probably faeces, but nonetheless should be followed carefully. Testing for faecal blood was negative but the blood level of haemoglobin was found to be low at 9.9g/dl.

Mr A.C. was seen again three weeks later complaining of increasing deep seated gluteal pain which was made worse by warmth and eased by cooling, defaecation and occasionally by micturition. He had had no rectal discharge and was passing two formed stools per day with no associated blood or mucous. On rectal examination a convincing extra-rectal pelvic mass was palpated. Mr A.C. was admitted and on examination under anaesthetic confirmed the presence of a hard, but mobile, mass approximately two by three centimetres in size, situated above the prostate gland in the
anterior rectal wall. On sigmoidoscopy a mild granularity of the mucosa was noted but no abnormalities in the vicinity of the mass were visualised. Histological examination of two true-cut needle biopsies from the mass revealed it to be a poorly differentiated, mucin secreting, adenocarcinoma lying apparently beneath the mucosa. No transition was noted into the mucosa, which exhibited changes of low grade chronic proctitis, nor into any other normal parenchyma. Acid phosphatase staining was negative ruling out the possibility of an extension of a prostatic neoplasm. A scan using computerised axial tomography confirmed the presence of a mass lying anterior to the rectum (Fig. 6) but no other abdominal abnormalities were noted. Gastroscopy was performed to exclude a silent primary gastric carcinoma with signet metastases. A number of small polyps (up to 2mm) were present in the fundus and body of the stomach which, histologically, proved to be purely metaplastic. It was decided to proceed to laparotomy and perform a panproctocolectomy and ileostomy if the mass proved to be resectable. However, at laparotomy the findings were free ascitic fluid along with extensive peritoneal and omental tumour deposits. The site of the primary tumour was not clear although the most likely candidate was a tumour mass on the lateral aspect of the mid ascending colon. There was no evidence of impending obstruction and with no therapeutic procedure practical the abdomen was closed.
Fig. 6. C.A.T. scan of the pelvis showing a mass lying anterior to the rectum.
DISCUSSION

All surveys have shown that there is an increased risk of colorectal carcinoma in ulcerative colitis, quoted as occurring at a rate approximately 20 to 30 times greater than that expected in a healthy population (Greenstein et al., 1981). There is general agreement that the tumours tend to occur at a younger age than in the general population and after a latent interval of about 10 years from the onset. The decade incidence of colorectal carcinoma in one series was reported to increase from 0.4% in the first decade to 7.4% in the second, 15.9% in the third and 52.6% in the fourth decade of follow-up. The estimated cumulative probability of developing carcinoma reached 34% at 30 years and 64% at 40 years (Greenstein et al., 1979). Although the accuracy of figures such as these has been disputed, not least because at present 20% of all patients with colitis are treated by proctocolectomy within the first 10 years, they are certainly significant and warrant management by surveillance and/or prophylactic surgery (Hendrickson et al., 1985).

One of the major clinical problems is that a significant percentage of ulcerative colitis patients present with tumours that have already spread beyond the bowel wall due primarily to difficulties in recognizing cancer in ulcerative colitis patients. The main reason for the difficulty being that the symptoms of cancer are very easily confused with the symptoms of the original inflammatory bowel disease and treated as such by the patients themselves. Thus, patients should be seen regularly whatever the symptomology being expressed. A recent study to assess the effectiveness of different forms of surveillance in the follow-up of 360 patients with mild to moderate symptoms of ulcerative colitis has just been completed (Lennard-Jones, 1985). Surveillance was not justified in patients with proctitis alone, a confirmation of the results of Greenstein et al. (1979), and relevant to the case described at initial
presentation. When barium enema suggests involvement of the transverse colon, Lennard-Jones recommends colonoscopy and biopsy to assess the extent of the disease. Recently Saverymillu et al., (1986) have supported the use of Induim granulocyte scanning and have reported excellent correlation with colonoscopy and histology. In this group of patients especially if young and in all patients with extensive disease (up to and including the hepatic flexure) regular check-ups should be made to supervise the colitis and maintain contact. Considerable discussion has occurred as regards the optimum time interval between appointments and the best methods of surveillance. Classically a barium enema and rectal biopsy are carried out every two years if the patient's condition remains relatively stable. Rectal biopsy is of value since dysplasia is thought to occur simultaneously at multiple sites along the colon since the whole mucosa is involved in the same inflammatory process (Morson and Pan, 1976). Riddell and Morson (1979) were able to detect 85-90% of III colorectal carcinomas on the basis of dysplastic changes in rectal biopsy specimens even though less than 50% of carcinomas were located in the rectum. However, in a more recent report rectal dysplasia was seen in only one of three patients with carcinomas and in only one of five carcinomas located in proximal parts of the colon (Vatn et al., 1984). Dysplasia was noted in the proximal colon in all cases. This study along with that of Riddell (1985) suggests regular colonoscopy, perhaps every two years, paying particular attention to the distal colon, together with general assessment, sigmoidoscopy and rectal biopsy in the intervening years. On the other hand there have been questions raised as to the value of colonoscopy; does the 'one biopsy every 10cm' rule, often employed, make sense if one biopsy is then thought to be representative of perhaps 100cm² of large bowel. Additionally, although dysplasia at remote sites has been noted retrospectively in all specimens of bowel resected for carcinoma, what is the risk of a patients with low grade or high grade dysplasia having or developing an occult (or even overt) carcinoma?
Colectomy should probably be recommended once unequivocal dysplasia (i.e. intra-epithelial neoplasia) is present. Although, again, other indications for prophylactic colectomy have been hotly debated, it is generally agreed that if the disease has been present for more than 10 years along with two or more of the following features, colectomy should be advised even if the patient appears to be in perfect health:

i) A severe first attack

ii) Radiological evidence of total involvement of the colon

iii) Continuous symptoms of colitis rather than intermittent attacks

iv) Onset of the disease in childhood or very early adult life.

The use of this policy resulted in only 3 cases of cancer developing in 700 patients attending the John Radcliffe Hospital between 1970 and 1980 (Lee and Truelove, 1980). However the patient described in our case did not convincingly fulfill these requirements and later presented with metastatic disease.

Whatever approach is used for making the decision to advise a prophylactic colectomy, the operation of choice is a proctocolectomy with formation of an ileostomy, although the recent revival of rectocolectomy with anal conservation has produced another less radical option for some patients.

Most carcinomas arising in the background of inflammatory bowel disease are difficult to diagnose because they tend to bear a greater resemblance to early gastric carcinoma than the usual appearances seen in early large bowel carcinomas. They are flat, plaque-like, or only slightly raised and are rarely ulcerated. Thus as in the case of the tumour of the ascending colon in the patient presented they may be very difficult to detect radiologically and even endoscopically. Further, even if visualised biopsy rarely shows invasive carcinoma although the overlying dysplastic epithelium is usually seen. Occasionally invasive carcinoma can be seen.
arising from a mucosa which shows no signs of dysplasia (Hendriksen et al., 1985), so that the rectal tumour seen in our patient may well be another primary. This multicentricity of large bowel tumours is a distinguishing feature from tumours in non colitis patients. Other features include the distribution of tumours, occurring at any point along the colon rather than predominantly in the rectosigmoid region, and the affected age group with a markedly increased incidence in patients under 40 (MacDermott, 1985).

The histopathological type of tumour often encountered, a particularly aggressive form of mucin-secreting adenocarcinoma, and the delay often incurred in detection of malignancy, due to difficulty in recognizing the symptomatic evidence of its presence, often means, unfortunately, as in this case, that patients are inoperable at presentation.
CONCLUSION

The results from this study have provided some interesting insights into the effects of...
CONCLUSION

The cases that have been described illustrate some of the many ways in which surgery may be employed in the management of patients with inflammatory bowel disease. Initially, laparotomy may be the only certain means of distinguishing Crohn's disease from a more sinister diagnosis such as lymphoma. Once a diagnosis of inflammatory bowel disease has been made some of the difficulties in differentiating between Crohn's disease and ulcerative colitis have been discussed along with the implications of each diagnosis for future management. Surgical treatment is often necessary for some of the many complications associated with these diseases, such as intestinal obstruction, perianal abscesses and anal fistulae. The great debates in the literature at present, however, involve the definitive excisional operative procedures. Most of the cases described demonstrate the difficulties involved in the decision to operate, each demanding that certain questions be answered. At what stage, if at all, in the acute and chronic forms of each disease should operation be performed and which type of procedure should be followed? Should prophylactic colectomy be performed routinely to eliminate the risk of carcinoma? Does chronic Crohn's colitis warrant intervention? If a prophylactic procedure is to be performed, at what stage in the illness and what criteria or method of surveillance should be used to determine this point? Although some attempt has been made to answer these questions drawing on evidence emerging from these six cases and from a review of the literature it is impossible to reach definitive conclusions. Treatment policies dependent upon the experience and opinion of individual surgeons continue to be the norm until deeper inroads have been made into the establishment of the aetiology and exact pathogenesis of the inflammatory bowel diseases. The tragic case of the patient who unexpectedly presented with inoperable carcinoma at the age of 41 superimposed on a history of mild stress related colitis confirms this general lack of understanding
of these diseases. The many cases of unexplained hepatobiliary disease, arthritis and perianal problems, along with the chronic misery of uncomplicated forms should fuel an active desire among the medical profession to uncover more of the hidden secrets of inflammatory bowel disease.

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