after survival of the acute illness is good (Thygesen, 1949; van Bogaert, 1950; Miller and Evans, 1953). That the ultimate prognosis of recovered neuromyelitis optica, which we regard as a variant of acute disseminated encephalomyelitis, is similarly favourable has been clearly shown by Scott (1952).

The differentiation of acute disseminated encephalomyelitis from acute disseminated sclerosis is therefore of much more than academic importance, but it may be very difficult in the acute stage. Both illnesses show signs of disseminated lesions in the neuraxis, both tend to occur in young adults, and both often follow non-specific infections, especially of the respiratory tract. In our experience the diagnostic criteria offered by McAlpine (1931) are invaluable, but the need for further study is demonstrated by the following recent case.

Seven weeks before admission to hospital a young man of 19 developed an acute influenzal illness, epidemic at the time in the Army camp in which he was serving, and associated in his case with quite severe pain in the left eye which lasted for three days. He recovered completely from his symptoms within a week, but seven days later vision in the left eye suddenly became blurred. Examination at this time revealed a unilateral retrobulbar neuritis with papillitis; there were no abnormal signs elsewhere. Sight in the left eye became normal within a week and the patient remained well until one month later, when he developed acute backache, followed by weakness and stiffness of the legs, constipation, and retention of urine. Examination at this time revealed some pallor of the left optic disk, nystagmus at the extremes of lateral gaze, and signs of a partial transverse lesion of the spinal cord in the mid-dorsal area, with loss of proprioceptive sensation in the feet and of impairment of superficial sensibility to the seventh thoracic dermatome. The spinal fluid contained 73 lymphocytes per c.mm., but was otherwise normal. Improvement from this lesion was rapid, and a mild spastic paraparesis with some left optic atrophy was at the time of writing the only evidence of his illness.

Was this a case of rapidly healing biphasic encephalomyelitis, showing some similarity to the syndrome of neuromyelitis optica, and with only slight danger of recurrence? Or must it be labelled as disseminated sclerosis, with all the sinister implications of that diagnosis? The answer to such problems seems less likely to depend on further refinements of clinical diagnosis than on the development of such ancillary aids as are suggested by the immunochemical studies of Kabat *et al.* (1948) or the refined colloidal gold reaction described by Lange and Harris (1949), which may ultimately furnish diagnostic instruments of critical importance.

Summary

Three further cases of acute disseminated encephalomyelitis personally treated with A.C.T.H. are reported, bringing the total of cases treated to 10. In seven patients marked improvement occurred during five-day courses of the drug, and in five of these unequivocal clinical improvement was observed within 12 hours of the initial injection of A.C.T.H. The two complete failures were in cases of subacute necrotic myelitis and catastrophic measles encephalitis. Improvement occurred within 12 hours in four of the six cases in which treatment was instituted within 48 hours of the onset of neurological symptoms, but in only one of four cases treated later in the course of the illness. This finding is regarded as a further indication that the course of the illness was favourably influenced by treatment.

Similar courses of A.C.T.H. were given during eight acute exacerbations of disseminated sclerosis in seven patients. Although euphoria was often present and improvement in chronic spasticity and bladder dysfunction occasionally observed (in keeping with results previously reported), acute symptoms showed no improvement comparable to that seen under similar conditions in acute disseminated encephalomyelitis.

The importance and the occasional difficulty of distinguishing between these two clinically similar but biologically distinct acute neurological syndromes are discussed and illustrated.

We wish to thank practitioner and consultant colleagues in Region No. 1 for their kind co-operation in referring cases for treatment, and the Medical Research Council for a supply of A.C.T.H.

REFERENCES

- Costero, I. (1949). *Arch. Neurol. Psychiat.*, Chicago, 62, 48.
 Fog, T. (1951). *Nord. Med.*, 46, 1742.
 Glaser, G. H., and Merritt, H. H. (1952). *J. Amer. med. Ass.*, 148, 898.
 Kabat, E. A., Glusman, M., and Knaub, V. (1948). *Amer. J. Med.*, 4, 653.
 Lange, C., and Harris, A. H. (1949). *Amer. J. clin. Path.*, 19, 16.
 Ligterink, J. A. T. (1951). *Ned. T. Geneesk.*, 95, 3490.
 Lumsden, C. E. (1951). *British Medical Journal*, 1, 1035.
 McAlpine, D. (1931). *Lancet*, 1, 846.
 Marburg, O. (1906). *Jb Psychiat. Neurol.*, 27, 213.
 — (1942). *J. Mt Sinai Hosp.*, 9, 640.
 Miller, H. G. (1953). *British Medical Journal*, 1, 177.
 — and Evans, M. J. (1953). *Quart. J. Med.* In press.
 Pette, H. (1942). *Die akute entzündliche Erkrankungen des Nervensystems*. Leipzig.
 Scott, G. I. (1952). *Amer. J. Ophthalm.*, 35, 755.
 Thygesen, P. (1949). *Arch. Neurol. Psychiat.*, Chicago, 61, 339.
 Van Bogaert, L. (1950). *J. Neuropath.*, 9, 219.
 — Borremans, Reusens, and Weyn, R. (1932). *Presse méd.*, 40, 1370.

CONSERVATIVE SURGERY IN THE TREATMENT OF ULCERATIVE COLITIS

BY

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There is no doubt that the modern improvements in ileostomy bag design have relieved an ileostomy of some of its terrors, but, in spite of this and the reports of many patients who have returned to most of their former activities, it would be idle and false to pretend that the prospect of an ileostomy for the rest of life is one that does not bring dismay and gloom to those of finer sensibility. Many patients for whom an ileostomy is contemplated or performed are in their youth and in their early married and home life, and few can believe that such distortion of the normal channels of evacuation is likely to prosper that alliance and that household. It is surely right that surgical opinion should not accept the inevitability of permanent ileostomy combined with total colectomy and excision of the rectum as the routine surgical treatment of the disease without the fullest exploration of more conservative methods. Devine and Devine (1948), in a series of cases, have put forward a technique whereby the rectum is preserved and continuity of the intestinal track re-established; and Corbett (1952), working along similar lines, has reported successful cases. Both of these methods entail the full discharge of ileal content through an ileostomy that is retained for varying periods of time, but the method of continuity suggested in this paper, dependent as it is upon immediate ileo-pelvi-rectal anastomosis, carries no such disadvantage.

Those antagonists of any attempt to preserve rectal function contend that the inflammatory changes present in the rectal mucosa continue in spite of removal of the rest of the colon and that the mucosa is unable to withstand the proteolytic effect of the ileal contents which are poured into it. Moreover, they regard the possibility of cancerous change in the remaining portion of the bowel as a very real one, and in the event of existing

complications of the disease being present, such as arthritis or pyoderma, they believe that these are not cured so long as a portion of the large bowel remains.

In certain cases, at least, it is doubtful whether these fears are real, and it is still more dubious whether they should prevent the surgeon from an attempt at restoration of continuity of the bowel. Devine and Devine have described the regression of the acute inflammatory change in the rectum, once its connexion with the colon has been severed, so that the mucosa returns practically to its original state, and my own experience supports this view. That the rectum and anal canal can withstand the proteolytic action of small-intestinal content has been proved to my satisfaction by my experience with anastomosis of the small intestine to the rectum for other conditions (Aylett, 1951, 1952), so that this consideration, too, cannot be held as a bar to attempts at functional restoration.

Suitable Cases

In the type of case upon which a conservative operation is proposed, the possibility of carcinomatous change in the remaining rectum is not a real one. Counsell and Dukes (1952) have shown that in ulcerative colitis cancerous changes occur either in association with a dense fibrous-looking stricture or as a result of malignant changes supervening in true adenomata. The common inflammatory pseudo-polyps and oedematous mucosal tags are not pre-malignant and may disappear completely with the amelioration of the disease. Where the rectum is studded with true adenomata the nature of which will be revealed by pre-operative biopsy examination, or where, in the course of sigmoidoscopic examination, stricture formation is identified in the portion of the bowel it is proposed to retain, any consideration of conservative surgery must naturally be abandoned in favour of an ileostomy with complete removal of the colon and rectum. In the absence of such changes, even though the typical acute inflammatory appearances with pseudo-polyps are obvious, I consider that the operation should be planned with a view to retention of the rectum.

Of the five cases reported below, one had a severe pyoderma. In this case at least, the condition resolved completely, and the return of the rectal mucosa to a normal appearance encourages us to believe that other extra-bowel lesions may resolve in a like manner, with a conservative operation retaining rectal function.

It is not within the scope of this paper to consider the numerous indications for surgical intervention in this disease, but if conservative surgery is to be contemplated it is essential that stricture formation in the rectum and the lowermost part of the colon shall be absent. In dealing with the case that is under prolonged medical treatment the closest co-operation between physician and surgeon is therefore essential, so that operation shall not be delayed until this complication has developed. It is only by sigmoidoscopic examination that the early stage of bowel changes likely to go on to stricture formation can be recognized, and this investigation must therefore be carried out at frequent intervals throughout the patient's illness. Distensibility of the rectum and colon is at all times diminished in cases of ulcerative colitis, as the normal elasticity of the bowel is reduced by the oedema and inflammatory changes that affect all its layers, but the start of actual stricture formation is suggested when the sigmoidoscope feels held up or gripped at any level of the bowel within the scope of the examination, or if the wall feels thickened and unduly resistant. In all such cases medical treatment should be abandoned in favour of surgery, as, if the former is prolonged, stricture formation will develop and retention of a functioning rectum will be impossible.

A conservative operation will also be out of the question when perirectal abscess and fistula formation is present,

but if the disease is regarded as one in which both physician and surgeon have an equal interest, and one in which operation does not carry the inevitability of permanent ileostomy, then the disease is unlikely to be allowed to reach the advanced stage before surgery is undertaken. Finally, the prospect of the retention of rectal function, by overcoming both the patient's disinclination to submit himself to an operation which is to result in a permanent ileostomy and the physician's natural reluctance to advise such a procedure until the very last, may help to avoid the "Belsen" type of case in which any operation must carry an appreciative mortality.

Operative Technique

The abdomen is opened through a left paramedian incision and the ileum divided close to the caecal valve. Both free ends are closed, and the proximal end is anastomosed to the pelvi-rectal region of the colon by side-to-side anastomosis.

The colon is then divided about 2 in. (5 cm.) above this site and the free ends are brought out on to the skin surface in the left iliac fossa (Fig. 1).

The patient is thus left with an end of the colon, a little above the anastomosis, opening on to the surface of the abdomen.

This provides a safety-valve effect, so that any pressure, the result of the accumulation of intestinal content or flatus in the rectum, which might be sufficient to break down the anastomosis between the ileum and the colon is prevented from developing, as the contents will discharge through the colostomy before enough tension to produce damage has been built up. In actual practice, although considerable quantities of flatus are passed, the amount of ileal content that escapes on to the abdominal surface is not great and has not been sufficient to produce excoriation of the skin. Nevertheless, I feel sure that the absence of any suggestion of a leakage in my cases at the anastomotic site has been due to the safety-valve effect of this colostomy, and that an intraperitoneal anastomosis carried out through grossly diseased bowel wall without this device would be fraught with great danger of leakage. The idea of decompression of the bowel by this method was first introduced by Muir (1947), who employed the technique when carrying out a right hemicolectomy.

In two of the five cases recorded below the condition of the patient at the completion of the ileo-pelvi-rectal anastomosis was good enough to enable the colectomy to be carried out at the same operation, so that only the distal free end of the colon was drawn out on to the surface of the abdomen. The general condition of most patients, however, would not permit of this, and the colectomy must be delayed until sufficient improvement has been achieved as a result of the ileocolostomy.

Closure of the safety valve some three to four weeks after the initial operation is the next step in the planned treatment of the condition. By this time the anastomosis will have united soundly, and there is then no need of the safety-valve effect. Moreover, it is best that this should be closed before the abdomen is reopened to remove the colon, as the risk of faecal contamination of the abdominal wound is thereby reduced. In Case 5 of the series reported, however, it was necessary to carry out the colectomy before the closure of the colostomy, as, after a period of great

FIG. 1.—The final altered anatomy, with the distal ends of the colon exteriorized.

improvement in the child's general condition following the first operation, deterioration began as a result of constant haemorrhage from the isolated colon. Colectomy was the final stage of operation in the remaining two patients. (Fig. 2.)

The progress of all patients after the ileocolostomy has been absolutely smooth and there has been no anxiety with regard to saline or fluid unbalance. Until a return of peristalsis, intravenous therapy has been continued, but, apart from an immediate post-operative blood transfusion, no continuance of this form of therapy has been necessary, even in those who, prior to the operation, needed weekly blood transfusions to maintain a normal blood picture.

FIG. 2.—The colectomy has been completed and the safety-valve colostomy closed. The photograph is that of a child aged 10 years.

The safety-valve colostomy discharges about the third day and ileus has never been severe enough to necessitate gastric suction. Once the bowels start to function, a day or two after the colostomy has acted, the discharge from this becomes steadily less.

The improvement in the general condition of the patient is rapid. Appetite and well-being return; fever and toxæmia disappear as quickly as after an ileostomy, and the emaciated patient puts on weight.

Case Reports

Case 1.—A woman aged 30, was admitted in November, 1951, with acute ulcerative colitis supervening upon a chronic condition of 19 years' standing. Bleeding and toxæmia were severe. In spite of all forms of treatment the condition deteriorated. Sigmoidoscopy showed appearances typical of an acute stage of the condition, with changes extending right down to the anal canal. The mucinase activity of the faeces was 400 units and the pre-operative weight was 6 st. 5 lb. (40.4 kg.). Ileo-pelvi-rectal anastomosis was carried out in February, 1952. The relief of symptoms and improvement in general condition were rapid. Colectomy was postponed until September, 1952. Before it was undertaken proctoscopy revealed a rectal mucosa that appeared normal. The ulcerative condition had completely disappeared. At the final operation the outward appearances of the distal segment of the colon and the upper rectum were seen to have changed dramatically. The sodden and inflamed bowel found at the initial operation had resolved and, with the exception of a slight thickening, appeared normal. The mucinase activity in the stool at the time of the final discharge was nil and her weight was 7 st. 11 lb. (49.4 kg.). The bowels were opened eight times in the 24 hours, and she was completely continent.

Case 2.—A man aged 46 was admitted in March, 1952, with a two-years history of ulcerative colitis. He had been in hospital elsewhere for the previous six months. Recently diffuse pyoderma of his face and neck had developed. Mucinase activity of the stool was 416 units. The bowels

were opened every two hours and there was a loss of weight of about 2 st. (13.2 kg.). His pre-operative weight was 8 st. 6 lb. (53.5 kg.). In view of failure to respond to medical treatment, operation was undertaken in April. Colectomy was carried out at the time of the pelvic anastomosis. Before discharge six weeks later the mucinase activity had fallen to 83 units. By September his bowels were being opened twice in the 24 hours, the pyoderma had subsided, and there was a weight gain of 17 lb. (7.7 kg.). The patient remains well. The ulceration of the rectal mucosa has healed entirely and with the exception of some oedema the mucosa appears normal.

Case 3.—A man aged 44 was admitted in March, 1952, with a seven-years history typical of ulcerative colitis. He had been in and out of hospital on numerous occasions, and in the acute attacks the rectal discharge necessitated visits to the toilet every 20 minutes. Sigmoidoscopy revealed changes of acute ulcerative colitis involving the rectum. The present attack failed to respond to medical treatment and anastomosis was carried out in May. The colectomy was completed in June. The patient's improvement was rapid. In September his bowels were opened eight to ten times a day and he was completely continent. He had gained 7 lb. (3.2 kg.) in weight, was fit and well, and was back at work. Rectal examination revealed a mucosa that was more oedematous than normal and which tended to bleed on pressure with the end of the proctoscope. The acute inflammation, with pseudo-polyposis formation present pre-operatively, had entirely disappeared. No blood was present in the stool.

Case 4.—A girl aged 10 was admitted in March, 1952, with a two-years history of periodic diarrhoea and blood in the stools. The haemoglobin on admission was 78%. Sigmoidoscopy and barium-enema examinations showed appearances of acute ulcerative colitis. The mucinase activity of the stool was 125 units. In spite of medical treatment the child's condition deteriorated steadily and multiple transfusions were required. At one time the haemoglobin fell rapidly to 46%. Blood-stained purulent discharge from the anus was almost constant, and operation was delayed only on account of the child's age. In September it seemed apparent that without operation she would die, and the anastomosis was therefore carried out. The improvement in her condition was dramatic. Three weeks later there was severe haemorrhage from the colostomy communicating with the isolated colon, and this continued. Colectomy was therefore carried out, and since that date her improvement to normal health has been uninterrupted. The mucinase activity of the stool is nil, and the rectal mucosa appears normal. The safety-valve colostomy has since been closed. There were six bowel actions in the 24 hours.

Case 5.—A woman aged 42 was admitted in July, 1952, with a 13-years history of chronic ulcerative colitis with acute exacerbations. She had been in hospital on numerous occasions. Recently there had been a steady decline in general health, constant blood-stained stools and discharge, and loss of weight. Sigmoidoscopy revealed typical changes of the condition down to the lower rectum, and barium-enema x-ray examination showed the terminal ileum to be involved. Ileo-pelvi-rectal anastomosis with colectomy and removal of the terminal 2 ft. (60 cm.) of small intestine was therefore undertaken. Since operation the general improvement has been great. The patient has put on 13 lb. (5.9 kg.) in weight and is in normal good health. She is completely continent, although the bowels are opened 10 times in the 24 hours. Blood is absent from the stool, and the pseudo-polyposis visualized and demonstrated by biopsy before operation has subsided.

Comment.—In the fulminating case of ulcerative colitis associated with perforation the same sphincter preservation should be aimed at. Such cases should be especially suitable for this type of operation, as the pathological changes occurring in the lower colon and rectum are acute and likely to be completely reversible. The general technique,

however, requires modification in view of the desperate condition of the patient. It is useless to attempt suture of the perforation, as the stitches will fail to hold in the diseased bowel wall, and, in any case, further perforations are usually impending at other sites in the colon. The caecum and colon as far down as the lower third of the pelvic colon must be excised, the distal end of the latter being brought out as a colostomy. Anastomosis of the ileum to the pelvi-rectal region would add a further quarter of an hour to the operation, and as the patient is unlikely to withstand this prolongation of operative time the severed end of the ileum is brought out as an ileostomy. Restoration of continuity of the bowel is delayed until the patient has fully recovered from the initial operation.

Discussion

I cannot agree with Counsell and Goligher (1952) in accepting the possibility of malignant change in ulcerative colitis as the dominating consideration in its surgical management. When true adenomata or stricture formation are present, then there is no question of the nature of the surgery that must be undertaken, but to advocate the removal of the rectum to prevent the formation of cancer when there is no certainty of its development is, I consider, too radical advice. The five specimens concerned in this article have been searched for cancer change and none has been found. It is unlikely to exist in the residua of the large bowels which have been retained, in view of the great improvement in the patients' general condition and the subsidence of the inflammatory changes, confirmed by proctoscopy. That the disease may recur in the rectum is a possibility, but the short follow-up of the present series has shown, so far, no indication of such a relapse. Even should a proportion have a return of symptoms, the establishment of an ileostomy and the excision of the rectum and terminal colon is no great surgical feat, and it can surely be reserved for those for whom it is a necessity rather than condemn all and sundry to a permanent ileostomy.

I have remarked on the noticeably smooth post-operative recovery of these patients, and from my own experience with ileostomies I have no doubt whatsoever about which is the easier. On this ground alone the operative mortality is likely to be very much less than the considerable mortality that accompanies ileostomy, and is an added argument in its favour.

Frequency of bowel action is unavoidable, but it is a small price to pay to escape an ileostomy life. In the second patient reported in the series the caecum appeared only mildly affected with the disease, and it, instead of the small intestine, was anastomosed to the pelvi-rectal junction. This patient has only two actions a day, and that this modification can be applied in other cases with advantage is a possibility.

Summary

The preservation of the rectum and of the normal channel of evacuation is advocated in those cases of ulcerative colitis in which fibrosis, pelvi-rectal suppuration, or true adenoma formation is absent from this part of the large intestine.

A method whereby this can be carried out without a preliminary ileostomy is described. The ileum is divided and its proximal end is anastomosed to the pelvi-rectal region. The colon above the site of the anastomosis is cut across and both free ends are exteriorized. The distal colostomy opening is subsequently closed and colectomy performed.

Immediate subtotal colectomy is advocated as the operative method of choice in the fulminating case.

Cases are reported in which a return to good health has followed treatment by the methods described.

I would like to thank Dr. E. R. Cullinan, in whose charge these patients have been, for his collaboration and advice in this work; Dr. C. B. Lewis, who has been responsible for the anaesthesia in these cases; and the nursing and resident staff of the Gordon Hospital for their unfailing care and attention to the patients.

Since this paper was prepared the initial stages of the operation have been successfully carried out in four other patients. One of these now has her bowels open three times a day only.

REFERENCES

- Aylett, S. O. (1951). *Proc. roy. Soc. Med.*, **44**, 196.
 — (1952). *Ibid.*, **45**, 394.
 Corbett, R. S. (1952). *Ann. roy. Coll. Surg. Engl.*, **10**, 21.
 Counsell, P. B., and Dukes, C. E. (1952). *Brit. J. Surg.*, **39**, 485.
 — and Goligher, J. C. (1952). *Lancet*, **2**, 1045.
 Devine, H., and Devine, J. (1948). *British Medical Journal*, **2**, 127.
 Muir, E. G. (1947). *Proc. roy. Soc. Med.*, **40**, 831.

PHLEGMASIA CAERULEA DOLENS

BY

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Phlegmasia caerulea dolens occurs in the limbs as a result of what would appear to be complete or almost complete obstruction to the venous outflow. It is of sudden onset, and is often associated with extreme cyanosis, swelling, pain, and shock, resulting in circulatory stasis of such a degree that gangrene follows in approximately a half of all cases.

The condition is rare, but has attracted much attention, particularly in France, and has been called variously pseudo-embolic phlebitis, blue phlebitis (Grégoire, 1938), and gangrenous thrombophlebitis (de Vega and Andrés García, 1946). DeBakey and Ochsner (1949) suggested the name phlegmasia caerulea dolens, and this seems an adequately descriptive term. Since then further cases have been reported (Clain and Nussbaum, 1950; Haimovici, 1950; Veal *et al.*, 1951; Miles, 1951; Turner, 1952).

Causation

The condition may occur when the main vein of a limb is suddenly obstructed. Veal *et al.* described cases that resulted from prophylactic ligation of the inferior vena cava in ilio-femoral thrombophlebitis with or without embolism, and I have seen the most alarming cyanosis and swelling after ligation of the common iliac vein for repeated embolism.

On account of repeated pulmonary embolism a man aged 63 had the common iliac vein ligated, thrombosis being present in the femoral vein. The following day he was moderately shocked and complained of severe pain in the thigh and leg. The affected leg was grossly swollen, intensely cyanosed, and much colder than his fellow, the distal pulses being hardly felt. The leg was elevated to 45 degrees and reflex heating and tolazoline hydrochloride were given; within three hours the swelling had practically disappeared and a full bounding pulsation was felt in the dorsalis pedis and posterior tibial vessels.

More usually it would seem to follow thrombophlebitis, occurring spontaneously or after operation, or in the puerperium, or as a result of pressure or invasion of a vein by neoplasm, or in association with blood disease.

A woman aged 54 was admitted to hospital with a thrombophlebitis of the right leg. She had suffered from attacks of phlebitis in the right leg during the thirty months prior to admission, and two and a half years previously had had a splenectomy, at another hospital, for a supposed Banti's syndrome. She was treated in hospital with penicillin, heparin, and dicoumarol, but, as a result of haemarthrosis and epistaxis, anticoagulants had to be stopped. Five weeks later there was a sudden onset of severe pain, swelling, and cyanosis of the right leg.