

# Familial adenomatous polyposis: the large bowel

**J P S Thomson DM MS FRCS**

Consultant Surgeon and Honorary Director

The Polyposis Registry, St Mark's Hospital, London

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For almost a century patients with polyposis, and the relationship of polyposis to cancer, have been studied extensively by those involved in their care at St Mark's Hospital. The first recorded operation for polyposis was carried out on 18 March 1918 and in 1925 the Polyposis Register, which has formed the database for all subsequent clinical and research activity, was started by H J R Bussey, the late Cuthbert Dukes and the late J P Lockhart-Mummery. It was not surprising, therefore, that the latter's son, H E Lockhart-Mummery had an interest in these important, hereditary disorders. This interest is recorded in six contributions made by him to the literature (1-6).

In 1966, Lockhart-Mummery (3) chose as the subject of his President's address to the Section of Proctology of The Royal Society of Medicine *Intestinal Polyposis: The Present Position*. In this address he emphasised the importance of recognising the varying types of polyposis and commented on several of these. It was, however, familial adenomatous polyposis (familial adenomatosis coli as he styled it) that received the most attention. This was entirely appropriate as this disorder is more common and better understood than some of the other forms of polyposis. Since that time, nearly 25 years ago, there have been changes in the management of the large intestine, but the fundamental principle of preventing large bowel malignancy by removing the colon and monitoring the rectum (colectomy with ileorectal anastomosis) remains. The added experience with this procedure to the end of 1989 is analysed with particular reference to postoperative mortality and morbidity, function and the risk of cancer in the rectum. It must now be remembered, though, that Familial Adenomatous Polyposis (FAP) is a systemic disorder which may affect the whole gastrointestinal tract and other tissues at other

sites and that patient management must take this into account, together with the clinical aspects of a rapidly expanding knowledge of its genetics.

## Which operation?

On 8 December 1948 the late O V Lloyd-Davies performed the first colectomy with an ileorectal anastomosis at St Mark's Hospital. This operation, although it requires very careful rectal surveillance postoperatively, was a great advance as the alternative was a proctocolectomy necessitating excision of the rectum and a permanent stoma. In the mid-1970s a further operation became available, namely restorative proctocolectomy. All three procedures have a place in the management of the large intestine as shown in the accompanying diagram (Fig. 1). The choice depends on the state of the rectum. In patients with masses of small adenomas which are quite uncontrollable, large sessile adenomas or malignancy in the upper or mid-rectum, a restorative proctocolectomy should be considered. Lockhart-Mummery referred in his paper in 1967 (3) to the problem of uncontrollable

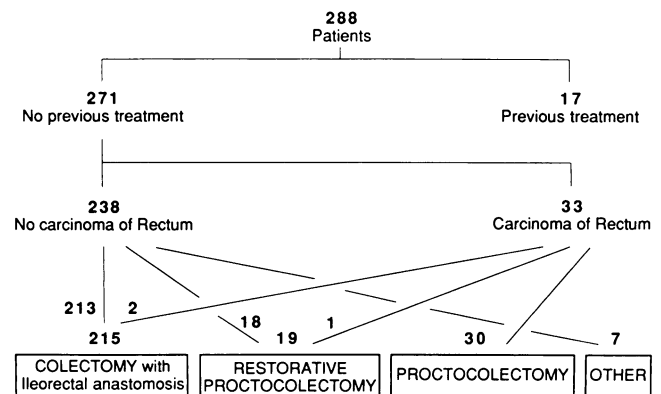


Figure 1. The surgical options in the St Mark's Hospital series of patients 1948-1989.

Correspondence to: Mr J P S Thomson, Consultant Surgeon and Honorary Director, The Polyposis Registry, St Mark's Hospital, City Road, London EC1V 2PS

polyps and his concern at having to excise the rectum for this. The surgical advance of restorative proctocolectomy, which owes so much to the late Sir Alan Parks, now provides an alternative. A proctocolectomy is probably only appropriate where there is a malignancy in the lower rectum (or perhaps in some patients the mid-rectum). It should have no place now in the management of entirely benign disease.

Colectomy with ileorectal anastomosis remains the first option in the management of the large intestine. In the last decade 85% of the patients at St Mark's have had this operation, 14% a restorative proctocolectomy and only 1% a proctocolectomy. It is also the preferred first option of the other members of the Leeds Castle Polyposis Group (7).

### Colectomy with ileorectal anastomosis

Lockhart-Mummery reported the first 65 patients who had a colectomy with ileorectal anastomosis at St Mark's Hospital. On 31 December 1989 this total had increased to 215 (Fig. 1).

#### Is the operation safe?

An uncomplicated postoperative course occurred in 193 (89.7%) of patients. The complications occurring in the remaining 22 patients were:

Anastomotic breakdown	5
Subphrenic abscess	1
Haemorrhage	2 (1 death)
Ileus/obstruction	14 (4 re-operation)

The only death occurred in 1953 and was a result of prolonged, intraperitoneal bleeding.

These current figures support Lockhart-Mummery's view that this was a safe procedure, but he did draw our attention to the prominent risk of intestinal obstruction. He analysed the complete series of St Mark's patients, that is, those who had a proctocolectomy as well as those who had a colectomy with ileorectal anastomosis and reported a very high incidence (20%) with a 3% mortality. It is not clear how many patients were operated upon. Our recent figures confirm a high incidence of an episode of obstruction but only one-half of these episodes required an operation to relieve it (Table I). The reason

Table I. Number of patients with postoperative intestinal obstruction (series total 214)

	Early (before discharge from hospital)	Late (after discharge from hospital)
Not requiring laparotomy	10	17
Requiring laparotomy	4	19
Total	14 (6.5%)	36*(16.8%)

\* There were, in fact, a total of 55 episodes of obstruction in these 36 patients. A total of 24 laparotomies were required (43.6%)

for this has not been determined but may be related to a disorder of fibrous tissue, prominent in those patients who have a desmoid tumour.

#### Is the outcome functionally acceptable?

In a previous study of a subgroup of 49 of the patients of this series (8) the mean bowel frequency was estimated to be  $3.4 \pm 1.7$  stools per 24 h. A more recent review in another subgroup of 61 patients (9) the mean bowel frequency has been calculated at 3.3 stools per 24 h. However, the range is 1–11 stools per 24 h with a median of 3; six patients having to rise at night for defaecation. Some patients may experience urgency, although 31 of the 61 patients (50%) can delay defaecation comfortably for more than 15 min.

#### How are the patients followed after operation?

The patients are seen at least once every 6 months and perhaps once every 3 months, especially if the rectum contains a large number of polyps. Lockhart-Mummery favoured the more frequent attendance (3).

At each visit the patients are assessed generally for any extracolonic manifestations of FAP. The rectum is carefully examined, after preparation with two slowly given phosphate enemas, in the jack-knife or knee-elbow position, with a large bore sigmoidoscope (1.7 cm external diameter). A suction apparatus facilitates this examination. Polyps may be readily diagnosed, although there may be difficulties in interpreting the findings (8).

With large numbers of patients attending, we have found it beneficial to have the patients come to specific clinics which are held every 3 months. Not only does this permit the above assessment but it facilitates arrangements for special investigations such as upper gastrointestinal endoscopy, research and also for screening those family members at risk.

Whilst sigmoidoscopy with biopsy is still the mainstay of assessment, colonoscopy with the dye-spray technique provides a very thorough and accurate examination of the whole of the large bowel. Assessment of congenital hypertrophy of the retinal pigment epithelium (CHRPE) and genetic testing (*vide infra*) now enhances our ability to establish whether or not a person at risk has FAP. Lockhart-Mummery was concerned about when to stop sigmoidoscopic assessment as he realised that neoplasms, benign and malignant, could arise at any time in those at risk. These newer methods of assessment allow much greater accuracy in diagnosis and will soon be adopted widely to define those who have the greater risk.

#### How are the rectal polyps managed?

Lockhart-Mummery changed our ideas about the management of the rectum (3). It had been traditional for the rectum to be cleared of polyps before undertaking the colectomy. This sometimes involved several admissions for diathermy fulguration. He drew our attention to the

possible theoretical risk of implantation of shed malignant cells from a carcinoma of the colon in the wounds created in the rectum by diathermy. He also drew our attention to the observation that after colectomy with an ileorectal anastomosis polyps do regress in size and number and may even disappear. This was observed in 1957 by Hubbard (10) and in 1959 by Cole and Holder (11) but it was not until 1988 that this was documented numerically (12).

For the last two decades, treatment of the rectum has been deferred until after the colectomy and usually only in those patients with polyps more than 5 mm in diameter. Diathermy fulguration has been the technique employed in all but a very few patients (snare removal or submucosal excision are other techniques). To date 124 patients (58%) have required treatment on 658 occasions. The sole complication which is rarely serious has been haemorrhage and this has occurred 17 times (2.6%).

**What is the risk of rectal carcinoma?**

It had been hoped that careful monitoring of the rectal stump and the appropriate destruction of adenomas would prevent the development of carcinoma. Unfortunately this has not entirely been the case, despite regular follow-up in the majority of patients.

There have now been 14 patients (6.5%) who have developed carcinoma. The follow-up compliance was good in nine patients and, of the remaining five, two were never seen after operation. Lockhart-Mummery reported the first two rectal carcinomas (3.1% of the 65 patients under review) in 1967. It has now been calculated that the cumulative risk of a rectal stump carcinoma developing is 10% at 25 years after colectomy (8). Reference to Fig. 2 shows that those developing carcinoma in the rectum have had the colectomy at a wide variety of ages. The youngest age at which carcinoma occurred was 28 years. The only factor identified which suggests a greater risk is the presence of a carcinoma in the resected colon.

All these patients with carcinoma have been treated by proctocolectomy with an ileostomy. It is noteworthy that the pathological stage was in some instances advanced—Dukes' A 6, Dukes' B 3, Dukes' C 5. Of the 14 patients, four eventually died from metastases.

Colectomy with ileorectal anastomosis is therefore a safe operation with a satisfactory functional result. It can confidently be recommended to young asymptomatic people as a prophylactic operation. There is a risk of rectal carcinoma and this necessitates careful follow-up. However, only four people have died from carcinoma of the rectum so far in this series of 214 patients; this is less than 2% using gross figures. The cumulative risk, however, is about 5% at 25 years.

**Restorative proctocolectomy**

The operation of restorative proctocolectomy has been performed in 19 patients in the St Mark's Hospital series (Fig. 1) and also in a further 17 patients. The results are yet to be published (9). There has been no mortality but a

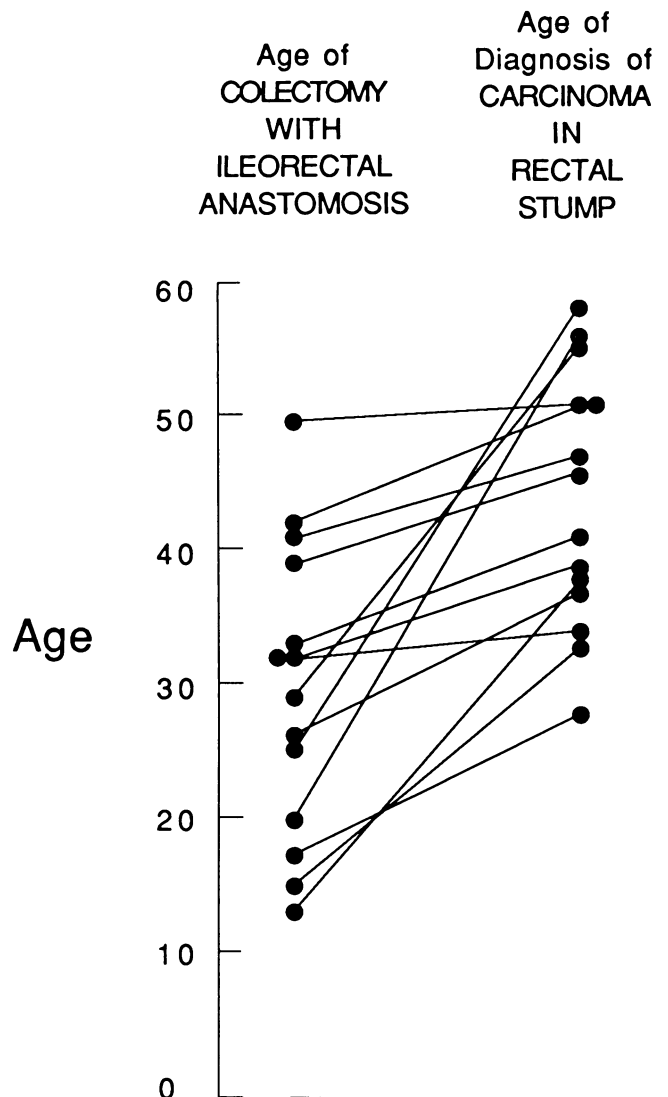


Figure 2. Carcinoma in the rectum after colectomy with ileorectal anastomosis—the age at operation and the age at presentation of carcinoma.

greater morbidity, mainly due to pelvic sepsis and complications related to the ileostomy. The functional results are very similar to those after colectomy with ileorectal anastomosis and, in fact, a smaller proportion of patients have urgency. Similar good functional results are also reported in a small series by Everett and Forty (13).

There is no doubt that this operation should be advised when appropriate, but its greater complexity, the possible need for a temporary ileostomy and increased morbidity suggests it is not an alternative to colectomy with an ileorectal anastomosis but only an alternative to proctocolectomy. In addition, the biological effect of creating a pouch on adenoma formation (and perhaps adenocarcinoma also) in the terminal ileum needs evaluation.

**Conclusion**

Sir Hugh Lockhart-Mummery had a great interest in familial adenomatous polyposis and while on the staff of

St Mark's Hospital he had the largest individual experience of this disease. He contributed to our knowledge and has helped to formulate our ideas. After 25 years his general principles for the management of the large intestine still hold and are supported by these satisfactory results.

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