

# **Three cases of Intestinal Polyposis in a family**

by

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Multiple polyposis or as American authors prefer to call familial intestinal polyposis is a rare familial condition characterized by a colon and rectum studded with multiple adenomas of all sizes and shapes including the flat or pedunculated varieties.

As the name denotes, it is a hereditary disease transmitted to both males and females. The affected patient is likely in turn to pass the condition over to half the children. Although hereditary, the disease usually lies dormant for sometime and manifests itself in childhood and most frequently in young adult life. Too often the onset of symptoms is insidious. The polyps give rise to diarrhea, excess passage of mucus, bleeding and abdominal pain. Important is the tendency of polyps to malignant degeneration.

So insidious may be the onset of the condition that for a long time the patient might accept the frequency of his bowel action as his normal habit, and the symptoms that make him seek medical advice would be those of carcinoma.

Diarrhea and blood loss by and by deteriorate the general condition of the patient and emaciation, severe anemia and cachexia supervene. So rapid may be the downward trend of disease and extensive development of carcinoma that radical treatment would be precluded.

A case of familial intestinal polyposis should make the physician to search for similar cases in the same family.

Diagnosis is made by sigmoidoscopy and x-ray (Barium enema). Proctoscopically the polyps situated in lower sigmoid and rectum are visualized. If no polyps can be found up to 20-30 cm. of anal verge, existence of the disease is ruled out. Fluoroscopically, a characteristic appearance is lent to colon owing to numerous small filling defects. Anyhow, x-ray after barium enema is always mandatory to confirm diagnosis and reveal the spread of polyps.

### **Treatment:**

Radical surgical excision of the involved colon offers the only choice of treatment. There are suggested three kinds of operations of which the author prefers the second and third procedures. Here are briefly described the three operations widely employed.

1. Ileostomy accompanied by total colectomy: As the artificial anus opening in iliac fossa is greatly abhorred in Iran we rarely choose this procedure. Dukes and Murrey of St. Mark's Hospital give an indication for this operation in all cases which malignant degeneration in rectum has supervened. The operation can be performed in one or two stages depending on the general condition of the patient.

### **2 Ileo-Rectal Anastomosis with total colectomy:**

This operation was performed in case II. It is a one-stage operation, extirpating the whole colon and preserving the rectum for end-to-end anastomosis with ileon. The operation is preceded by clearance of polyps from rectum which is performed in several sessions with diathermy. Close contact with these patients must continue for a long time after operation as there is a tendency for rectal polyps to recur. In such case the newly formed polyps should be coagulated immediately. The result is satisfactory and the bowel movement is reduced to two to four times in 24 hours.

### **3. Ileo-Anal Anastomosis with total Proctocolectomy:**

This operation is total colectomy and amputation of rectum supplemented by ileo-anal anastomosis. The whole procedure is performed in one-stage operation. Anal sphincter is preserved. Case III, underwent this operation.

### Case Reports

The three following cases occurred in a family with apparently no previous history of the affection as far as we could investigate. Two were brothers of 8 and 7 years old at the time of seeking medical advice and the third one was their sister of six.

Concerning the first case, the parents refused the proposal for surgical operation and in this way lost the oldest son within a short time. But this was a dear price to pay for their ignorance so that when the same disease affected the two other children they no more hesita-

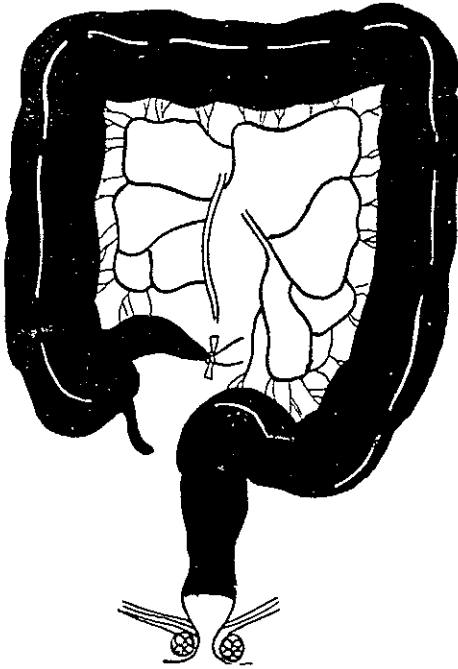


Fig. 1

Amount of colon removed in  
Case II.

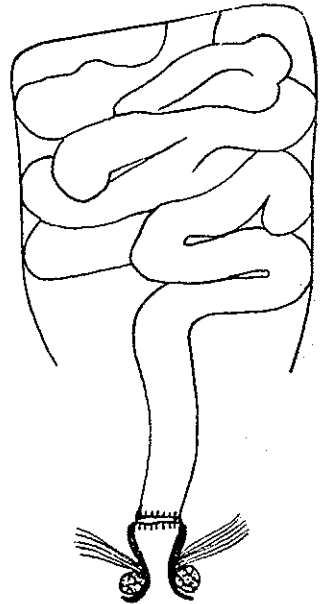


Fig. 2

Shema of operation performed  
in case II.

ted to accept surgical treatment. The patients underwent surgical operation satisfactorily and are in a good general condition since operation.

#### Case I

M. T., an eight years old boy was admitted to hospital in January 1948. The child's chief complaint was the frequency of bowel movement associated with bleeding and mucus discharge for the last four months. Rectal touch revealed the presence of numerous polyps studding the mucosa.

Sigmoidoscopy confirmed the diagnosis of multiple polyposis and X-ray of colon showed the characteristic appearance of numerous small filling defects all over the colon from cœcum to rectum. The parents refused operation. Dehydration and cachexia undermined his health within a short time. Repeated transfusions, administration of vitamins and liver extract, and protein therapy were of no avail. After  $1\frac{1}{2}$  year the patient died.

### Case II.

In September 1950 the six years old sister of the aforementioned patient was referred to hospital. Her complaints were the same as her brother's. So the diagnosis was not difficult. Rectal touch, sigmoidoscopy and X-ray proved the condition to be multiple polyposis. The parents had learned a bitter lesson already, so the proposition for total colectomy was accepted.

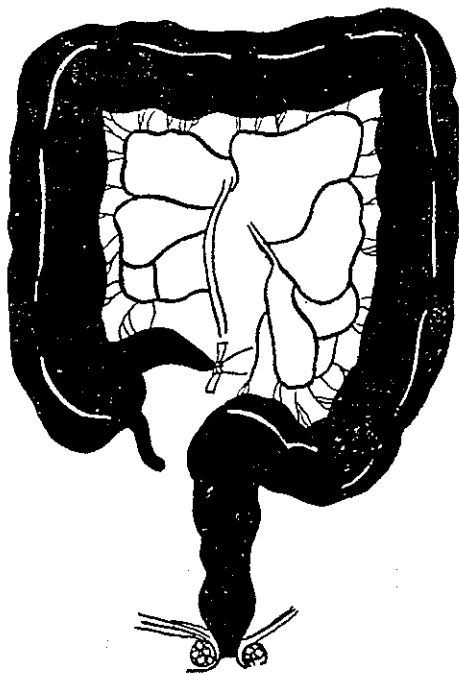


fig. 3

Amount of colon removed  
in case III

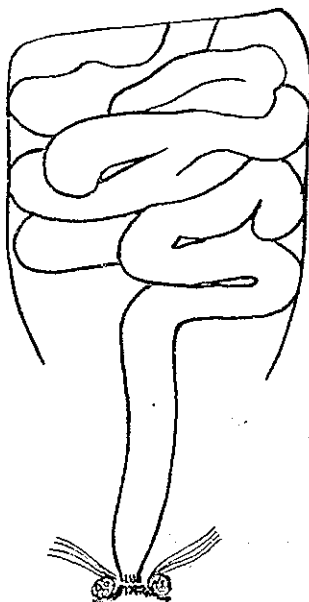


Fig. 4

Schema of operation performed  
in case III

**Surgical Operation:** Preceding the operation rectal polyps were removed with diathermy through a large bore sigmoidoscope in several sessions. The operation consisted that of a total colectomy through a median abdominal incision and end-to-end anastomosis of ileon to rectal

stump A drainage tube directed to lower portion of pelvis was inserted into peritoneal cavity and the abdominal wall closed.

After four days the drain produced what seemed to be bowel content. Fecal fistula had been established. The fistula did not close after a month of expectant therapy. So the patient underwent another operation and the fistula healed. For two years the patient's condition was quite satisfactory, when rectal bleeding began again. Sigmoidoscopy revealed the formation of new polyps on the remaining rectal mucosa. The new polyps were removed by coagulation. Since that time rectal function is quite satisfactory. The stools are soft to some extent. (Figs. 1, 2)

### Case III

The seven years old brother of the former patient was referred

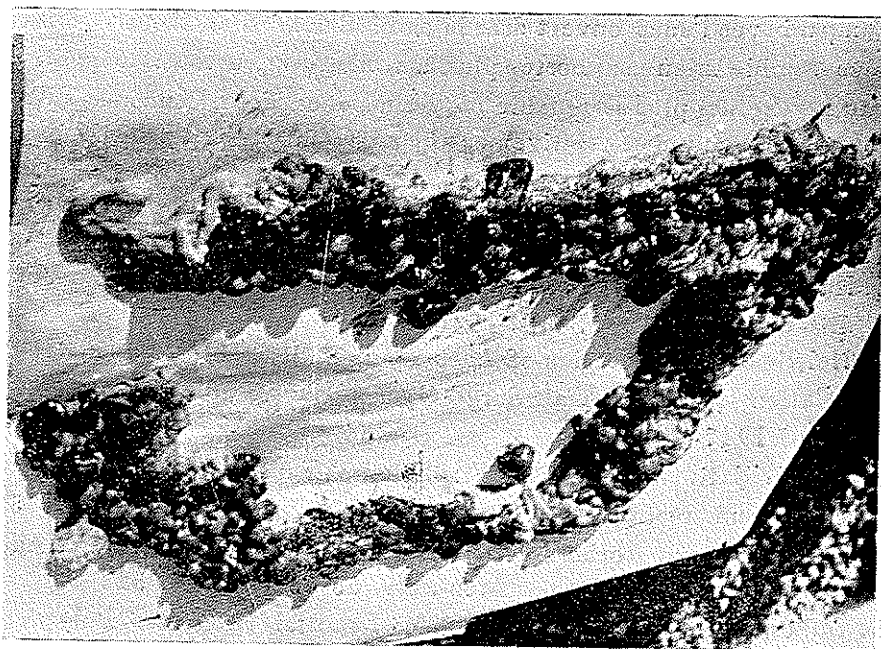


Fig. 5  
Specimen removed in case III.

to hospital in September 1955. Diarrhea and rectal bleeding were his complaints. Sigmoidoscopy and x-ray confirmed our diagnosis of multiple polyposis.

Surgical operation: Through a median incision the large bowel was removed. Rectum was extirpated trans-perineally, and the proximal end of ileon passed through. Pelvis was sutured to anal skin fold. The patient left the hospital after fifteen days of quiet convalescence.

Since operation the patient's condition has been quite favorable with no complaint of bowel movements. (Figs. 3,4,5)

### Trois cas de Polypose intestinal dans une famille

Les auteurs rapportent l'observation d'une polypose recto-colique généralisée familiale ayant atteint 3 enfants d'une même famille.

Le premier n'a pas été opéré malgré l'insistance du chirurgien. Il est mort à l'âge de 12 ans de l'anémie et des infections intercurrentes.

Pour les deux autres nous avons réussi à faire une coléctomie totale en un temps avec l'abaissement de l'iléon jusqu'au périnée. Comparé avec l'anus ouvert dans la paroi abdominale, l'anus périnéal nous semble mieux supporté, plus continent, et surtout, au point de vue psychique, plus acceptable par les malades. Actuellement ces deux enfants sont en vie et relativement bien portant (l'un est opéré il y a déjà six ans, l'autre un an et demi).

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