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CASE REPORT

Is STEP the future for patients requiring proctocolectomy? A new therapeutic proposal from pediatric experience

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Key Clinical Message

We present a pediatric case of medically unmanageable juvenile colonic polyposis, initially treated with subtotal colectomy and an ileostomy followed by a proctectomy, ileal-J-pouch and serial transverse enteroplasties (STEP) of the distal ileum. The STEP procedure in an adequate length was able to control stooling of our patient.

Keywords

Juvenile polyposis coli, paediatric, proctocolectomy, serial transverse enteroplasties, serial transverse enteroplasty.

Introduction

Certain colorectal conditions, such as ulcerative colitis and diffuse polyposis of the colon and rectum, and congenital intestinal abnormalities, such as total colonic aganglionosis, require a proctocolectomy and some type of intestinal restoration [1]. Although these procedures are technically demanding for surgeons, they are more challenging for the patients following the restoration of intestinal continuity. Frequency of bowel evacuations typically increases dramatically postsurgery, contributing to induced metabolic derangements [2, 3]. Several methods have been used to improve this morbidity with inconsistent results. We present a case of juvenile polyposis coli, initially treated with a subtotal colectomy and ileostomy, followed by a proctectomy, ileal-J-pouch and serial transverse enteroplasty (STEP) of the distal ileum [4].

Clinical case

An eight-year-old female patient was referred to our department with a chronic history of unremitting bloody diarrhea, weight loss, anemia and intractable rectal prolapse. Reduction of the rectal prolapse was always easily achieved by the mother. There was no other significant past medical history or family history and her vaccinations were up to date. The initial clinical examination revealed a pale child who was underweight for age with mild clubbing of the fingernails. The abdominal examina-

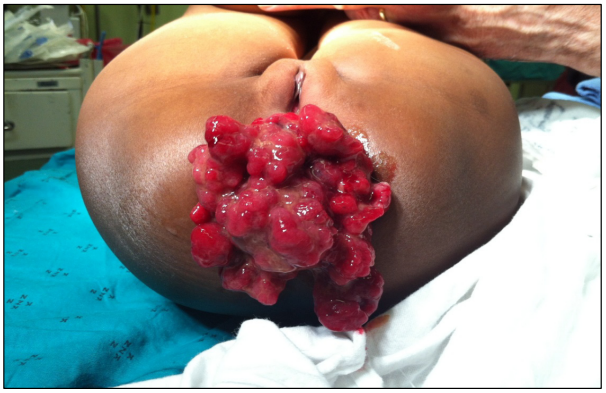

Figure 1. Prolapsed rectum demonstrating rectal polyposis.

tion was unremarkable, and the rectal examination was unnecessary because the cause of the prolapsed rectum was obvious, with a presentation of multiple polyps (Fig 1).

Blood tests confirmed clinical evident anemia with a hemoglobin of 7 g/dL, and her albumin level was 19 g/L. Rectal polyposis was ratified by sight (Fig 1), and total colonic involvement was suspected on the contrast enema (Fig 2) and was substantiated by colonoscopy (Fig 3) and by the subtotal colectomy specimen (Fig 4). The histological diagnosis of hamartomatous juvenile polyposis was confirmed from the colonoscopic biopsies and the operative specimen.

Despite the initial temporizing approach of subtotal colectomy and ileostomy, anemia and protein-losing enteropathy persisted. The patient was discharged with dietary supplementation, and she had regular outpatient follow-up. The child's nutritional status improved over time, and she had albumin levels consistently over 36 g/L. Three months after her initial surgery, she underwent definitive surgery. A proctectomy was performed to remove the residual rectal polyps, and the ileostomy was taken down and fashioned as a J-pouch (Fig 5). Multiple serial enteroplasties (Fig 6) starting just above the pouch over a length of approximately 80 cm was performed, followed by a hand sewn ileo-anal anastomosis.

The postoperative course was uneventful, and the patient was discharged 2 weeks later. At that stage, she was passing two soft stools per day without any medication or dietary restrictions. The follow-up at 3 months postsurgery confirmed a well-child who had gained 1.5 kg. She had returned to school and continued to pass one to two soft stools per day.

Discussion

Restorative proctocolectomy has replaced disparate surgical options such as a Hartmann's procedure and permanent ileostomy or ileo-rectal anastomoses because of their techni-

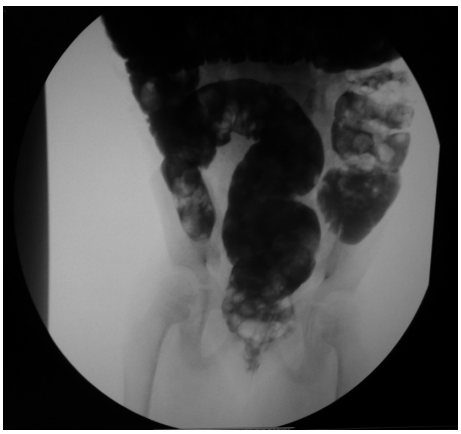

Figure 2. Contrast enema demonstrating multiple colonic mucosa defects suspicious of total colo-rectal polyposis.

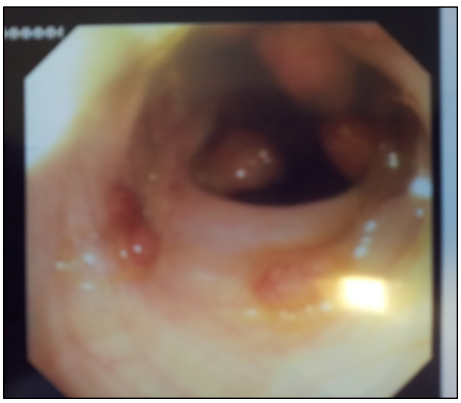

Figure 3. Polyps observed at colonoscopy.

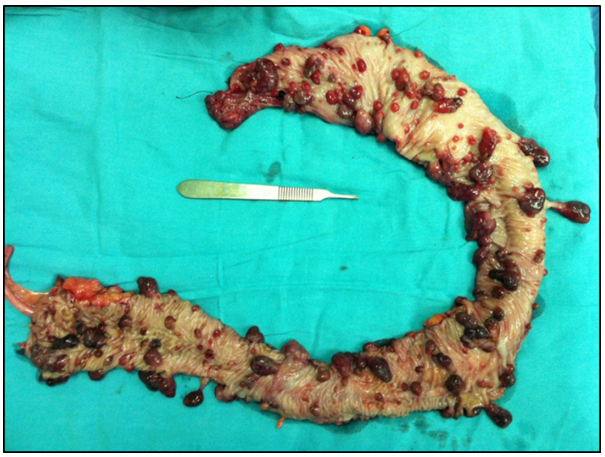

Figure 4. Subtotal colectomy specimen demonstrating the entire colonic mucosa colonised by polyps.

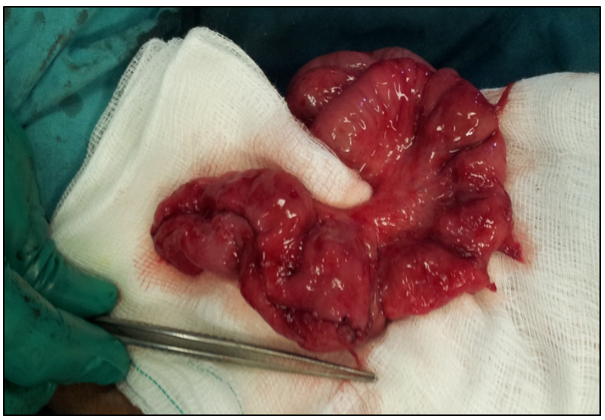

Figure 5. Ileal J pouch.

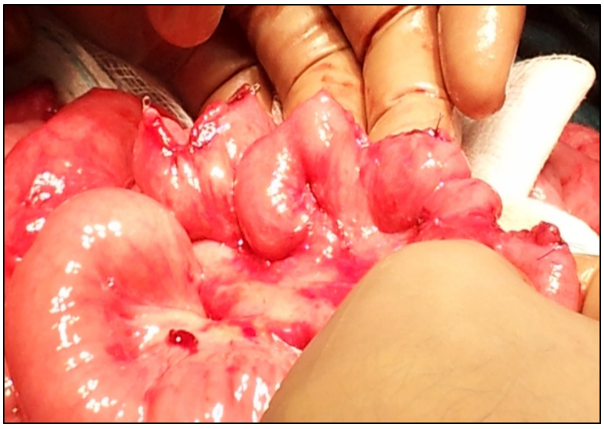

Figure 6. Serial transverse enteroplasties.

cal inadequacy, inability to improve quality of life and persistent cancer risk [5, 6]. Proctocolectomy is the surgical procedure of choice. It removes the diseased intestine, thereby improving quality of life and reducing cancer risk [3]. Intestinal leaks and pouchitis are major morbidities that might result in a long term or permanent ileostomy. An increased frequency and volume of enzyme-rich liquid stools may challenge the patient's continence and lead to severe perianal excoriations [6]. Although dietary restrictions and opioids might temporarily reduce the symptoms, they fail to cure these patients [7, 8]. In these cases, the postoperative quality of life is indisputably not better than the pre-operative status.

It is well reported that patients affected by severe short bowel syndrome benefit from the STEP procedure, because such an intervention can significantly reduce the frequency of stooling [9–13]. Proctocolectomy deprives the patient of their "constipating" intestine, leading to diarrhea. Such large volumes must therefore have time and an appropriate intestinal length for water, electrolytes and other nutrients to be absorbed. This process is expected to occur within the J or other pouch configurations following restoration [14, 15]. These pouches are quickly filled, and they easily surge; the anal sphincter does not effectively counteract and contain the volume, which is usually controlled by the ileocaecal valve. The brisk small bowel peristalsis ensures that the situation is even more pressing. Large amounts of nutrients, electrolytes and water are wasted because evacuation is inevitable and continence training is not possible. The patients homeostasis rapidly deteriorates if such losses are not promptly replaced. STEP is a technique for lengthening the intestine in short bowel syndrome by interrupting the intestinal lumen thereby also slowing peristalsis. This procedure allows time for the nutrients, water and electrolytes contained within the intestinal lumen (and distant from the distal anal anastomosis) to be absorbed [4, 11].

The outcome of our technical option was rewarding because the stool frequency and consistency were immediately in an acceptable range of two to three times per day and a soft consistency, respectively. This outcome was achieved without the use of opioids or the need for dietary restrictions. The child tolerated an unrestricted hospital diet and was discharged after 2 weeks when we were certain of her bowel habits and that she had maintained her physiological parameters. At 3 months follow-up, she was growing well and maintained a stool frequency of one to two soft stools per day. Stool frequency can be crippling following proctocolectomy, and although opioids might provide temporary relief of the symptoms, they cannot prevent relapses. We believe that STEP alone, performed in an adequate length of bowel, can control the stooling of such patients.

Conflict of Interest

None declared.

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