SURGERY FOR INFLAMMATORY BOWEL DISEASE IN CHILDREN AND ADOLESCENTS

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Recent decades have seen a constant rise in the incidence of IBD in both adults and children. Despite considerable progress in the pharmacological treatment of this disease, surgery has become the more frequently used treatment modality in younger patients. In the presence of massive haemorrhage, free perforation, fulminate colitis or acute obstruction, only surgical intervention has a chance of saving the patient’s life.

The aim of the study was to present the results of surgical treatment of IBD in children and adolescents who were operated on in a department which copes with “adult surgery” in its everyday practice.

Materials and methods. 235 patients were operated on for IBD in the years 1998-2005. There were 18 (7,66%) children in this group, 10 girls and 8 boys. 12 patients were diagnosed with ulcerative colitis (66.7 %) and (6) patients were diagnosed with Crohn’s disease (33.3%). The age of the patients ranged from 12 to 17 years (mean 15.6). Among the 18 children, 10 (55.6%) were operated on for elective reasons and 8 (44.4%) of the interventions were emergencies (three perforations, two obstructions, one acute haemorrhage and one fulminate colitis). In all cases of ulcerative colitis, a two-step restorative proctocolectomy with J pouch anal anastomosis was performed. Patients with Crohn’s disease were treated by limited (sparing) bowel resection and/or strictureplasty.

Results. There were no postoperative deaths in the study group. Postoperative complications were observed in 6 (33.3%) patients, the complications were ileus in 3 patients (1 patient demanded relaparotomy), pneumonia in 2 patients and wound suppuration with subsequent dehiscence in 1 patient. In one patient treated preoperatively with large doses of Imuran, the postoperative histology revealed a malignant lymphoma. Hospital stays ranged from 8 to 19 days (mean 12 days).

Conclusions. Surgery for IBD in children and adolescents has become a widely accepted method, and it is often the only treatment modality that offers a chance of a cure. Restorative proctocolectomy should be considered earlier in many cases of younger patients with ulcerative colitis, prior to conservative treatment, as immunosuppression and steroid therapy in particular produce undesired side effects. A consulting surgeon should be involved in the treatment of younger patients with IBD at a much earlier stage of therapy than is currently practiced.

Key words: ulcerative colitis, Crohn’s disease, surgery

Until recently, it was believed that inflammatory bowel diseases (IBD), which include ulcerative colitis (UC) and Crohn’s disease (CD), affect children less frequently than adults. However, it is currently recognized that in 20-30% of patients, the onset of the disease oc-
surgery in childhood or early adolescence before sexual maturity has been reached (1, 2). Additionally, a constant rise in the incidence of IBD has been recorded both in adults and in children (3). At the same time as surgeons are becoming more experienced in the treatment of IBD in adults, surgical intervention is more often performed as the method of choice in pediatric patients. In cases of massive haemorrhage, bowel perforation, fulminate colitis, toxic megacolon or acute bowel obstruction, surgery is the only treatment which offers a chance of saving the child’s life (4).

The aim of this study was to present the results of surgical treatment for IBD in children in our surgical department, which routinely treats adult patients.

MATERIAL AND METHODS

The group of 235 patients were operated on for IBD between 1998 and 2005 at the Department of General, Gastroenterological and Endocrine Surgery in Poznań, Poland. There were 18 children in this group (7.66%), 10 girls (55.6%) and 8 boys (44.4%). Of the children, 12 suffered from ulcerative colitis (66.7%) and 6 from Crohn’s disease (33.3%). The age of the paediatric patients ranged from 12 to 17 years (mean 15.6).

Common presenting symptoms included abdominal pains in 18 patients (100%), diarrhoea (more than 4 stools per day) in 14 patients (77.6%) and fresh blood in stools in 11 patients (61.1%). Menstruation abnormalities were observed in all female patients. The time between the onset of the disease and the operation ranged from 3 months to 6 years (mean 2.3 years). 10 patients were on steroids before the operation and 3 received Imuran (Azathioprine). Symptoms of iatrogenic Cushing syndrome were present in 6 of the 10 patients on steroid therapy. Preoperative serum protein levels ranged from 3.8 g/dL to 6.7 g/dL (mean 5.1 g/dL). The patients’ absence from school because of symptomatic disease requiring hospitalization ranged from 3 to 25 months (mean 5 months).

RESULTS

In the group of paediatric patients, 10 operations were performed for elective indications and 8 for urgent or emergent reasons (3 ileus cases, 2 perforation cases, 1 severe bleeding case, 1 case of toxic dilatation of the colon and 1 giant retroperitoneal abscess with severe systemic septic complications).

In all UC patients, the first stage of surgical treatment consisted of a restorative proctocolectomy with ileal “J” pouch-anal anastomosis and a temporary loop ileostomy. This technique is currently advocated by both American and European centres renowned for their expertise in the surgical treatment of IBD (5, 6, 7).

In three CD patients, right hemicolectomies including the distal portion of the terminal ileum were performed. In two patients, small segmental bowel resections were carried out. In one child, the resection of 20 cm of the strictured ileum was performed together with a Heineke-Mikulicz strictureplasty of a 4 cm long narrowing of another portion of the ileum and a by-pass anastomosis of a nonresectable inflammatory tumour.

In five patients with CD, intestinal anastomoses were constructed with linear staplers, creating wide side-to-side anastomoses. The choice of this type of anastomosis was based on our own encouraging experience in adult patients and convincing positive data on this technique from the literature (8, 9, 10).

No postoperative deaths were recorded in the study group. Postoperative complications were seen in 6 patients (33.3%). Postoperative ileus occurred in 3 patients (16.7%), which in one case demanded relaparotomy. Pneumonia was observed in two patients (11.1%) and wound suppuration with subsequent dehiscence was detected in one patient (5.6%).

In one female patient treated preoperatively with large doses of Azathioprine (200 mg per day), histological evaluation of the resected colon revealed lymphoma.

The patients’ hospital stay ranged from 8 to 19 days (mean 12 days).

In all patients subjected to restorative proctocolectomy (n=12), the loop ileostomy was reversed in the second stage of surgical treatment. The follow-up period after ileostomy closure ranged from 6 months to 6 years. Functional results after the restoration of intestinal continuity were satisfactory. The patients had 2 to 7 bowel openings per day (mean 3.2) and only two patients reported night soiling of the underwear (16.7%). Clinical signs of pouchitis occurred in two patients (16.7%) and
were successfully treated with metronidazole and antibiotics. In one patient operated on for CD, the active disease relapsed, calling for reoperation (7.14%).

In February 2006, a 17 year old male patient was readmitted to the ward who had been operated on 17 months earlier for UC (a restorative proctocolectomy had been performed). Four months after ileostomy closure, the boy presented with severe symptoms of pouchitis refractory to all conservative measures. The patient was operated on; the pouch was excised and a terminal ileostomy was performed. Histological examination of the pouch revealed profound inflammatory changes typical of CD.

DISCUSSION

Ulcerative colitis and Crohn’s disease constitute a diverse group of clinical entities commonly referred to as non-specific inflammatory bowel diseases (IBDs). IBDs are increasingly affecting children and adolescents, and their presenting symptoms and complications make the choice of adequate treatment of paramount importance in paediatric patients. One of the most relevant issues is the timing of surgical intervention and the extent of surgery which will result in the best long-term results. Treatment of IBD in children demands attention to different factors than in adults. The evaluation of a child suffering from IBD should not be limited to intestinal symptoms. Extraintestinal manifestations are extremely important in qualifying a child for surgery. Growth retardation, delayed pubescence and psychosocial problems are extremely important in the paediatric patient group. The differences between adults and children imply the necessity of a varied approach to both conservative and surgical treatment. Kirschner (10) points out a lack of prospective trials on the efficacy of the conventional doses of non-steroidal anti-inflammatory medications, steroids and immunosuppressants, in particular age groups. He stresses the need for sufficient nutrition therapy augmenting the child’s growth and draws attention to the undesired side-effects of prolonged conservative treatment by medication, especially for steroids and immunosuppression.

Because of the different nature of these two diseases, the surgical treatment approach must also be based on different principles. In the case of CD, an operation is necessary in cases of severe complications. Indications for surgery include mechanical obstruction, septic complications (abscess, inflammatory tumour, fistula connecting the intestine to the urinary tract or resulting in short bowel syndrome) and free perforation. Additionally, an operation should be considered in all patients with severe, persistent anaemia resulting from protracted intestinal bleeding and in patients with grossly impaired intestinal absorption which produces cachexia or growth retardation (12).

In children, important elective indications for surgery include the failure of adequate conservative measures, fast relapse of the disease after steroid dose reduction, a lack of therapeutic effect at maximal doses, undesired side-effects or growing steroid-dependency. This view is shared also by El-Baba, who evaluated indications for surgery and long-term results. In his paper, he reported on 32 children of whom 22 were histologically diagnosed as CD. He listed the indications for surgery in the order of frequency of incidence: side-effects of pharmacotherapy, growth retardation, mechanical obstruction, septic complications and perforations. In 47% of the patients in this group, surgical intervention led to considerable alleviation of symptoms in the year following surgery. In 22% of the patients, active disease recurred, however the relapses were controlled much better with conservative measures. Two patients demanded relaparotomy and parenteral nutrition. The authors concluded that surgery considerably improves the patients’ quality of life, and best results should be anticipated in patients with locally limited disease (6). The decision of whether to operate for complicated cases of CD is still not an easy task, which was also confirmed in the study by Baldassano. The author reported that recurrences occur in 17% of patients a year after surgery, 38% after three years and approximately 60% after 5 years. He observed that in patients with disease limited to the colon, the time of remission ranged from 1 to 2 years. When the disease affected the ileo-caecal junction, the disease recurred after a mean of 4.4 years. Cases with multifocal localization of the disease [reached a] mean of three years without postoperative recurrence. The conclusion from his study was that recurrence is more frequent in children with more advanced disease and with disease localized in the colon. Resection of the affected portion of the bowel im-
proves the general condition and allows for normal growth of the child (14).

In this study, the technique of strictureplasty was applied in only one patient. The potential benefits of this method have been presented by Oliva among others. He reported strictureplasty (with or without bowel resection) in paediatric patients with multifocal localization of CD, and found considerably decreased intestinal symptoms resulting in a body mass gain and a marked reduction of steroid doses (15).

The philosophy behind the surgical approach to UC treatment is much different from that practiced in CD. In cases of emergent intervention, the procedure of choice is Hartmann’s colectomy with an end ileostomy and rectal remnant reaching above the peritoneal reflection. In cases of elective operations, total restorative proctocolectomy removes the primary site of inflammation and the potential site of severe dysplasia and subsequent malignancy. In the 1970s, attempts were made to replace the traditional ileostomy with an ileal pouch with a valve (Kock’s pouch). In 1987, Ein reported on his almost 10 years of experience in performing this technically demanding and complication-plagued surgery in paediatric patients. 15 months after pouch formation, no major problems were recorded. When pouch emptying was analysed, difficulties were detected, probably related to valve insufficiency. Major inflammatory changes in the pouch were observed between the 15th month and 5th year after pouch formation, while after five years the risk of any complications was almost none (5). However, except for papers by Kock and some of his followers, there is no convincing evidence proving this relatively complicated method to be superior to properly constructed (according to Brooke’s principles) end ileostomy (16). The necessity of pouch emptying through catheterization poses a serious problem, especially in the case of paediatric patients. According to the opinions of many patients, pouch catheterization and the associated necessary irrigations significantly decreased their quality of life.

In the cases when the inflammatory changes typical of UC are less prevalent or absent in the rectum, it is possible to complete a colectomy with ileo-rectal anastomosis. This procedure leads to good functional results, decreases the number of stools to a mean of 3 per day and lowers the risk of impaired continence. However, this is not the optimal technique to be used in children because of the necessary endoscopic surveillance of the left portion of the rectum. In that case, rectoscopy is mandatory in order to prevent severe inflammation, dysplasia and subsequent neoplastic transformation of the rectal stump.

Presently, the preferred surgical option for treatment of UC is restorative proctocolectomy. Paediatric surgeons often restore intestinal continuity after colectomy using Soave’s procedure. During this operation also known as the “pull-through” procedure, the terminal ileum is pulled through a rectal cuff that has been subjected to mucosectomy to the level of the dentate line. The introduction of an ileal pouch anastomosed to the anus just above the dentate line with preservation of sphincter function was a breakthrough in colorectal surgery. Even though restorative proctocolectomy has become a “gold standard” in adult surgery, the “pull-through” operation is still preferred by paediatric surgeons. The study published by Rintala confirms the observation that a restorative proctocolectomy with IPAA may be used in children with the same rate of success as in adults. Complications after restorative proctocolectomy are mainly related to preoperative therapy with immunosuppressants. The main problem directly associated with the underlying inflammatory condition is inflammation of the pouch mucosa (pouchitis), which may lead in extreme situations to pouch failure (6). Alexander published the results of his observations on a group of 151 paediatric patients after restorative proctocolectomy, with a postoperative follow-up period ranging from 2 to 15 years. He identified Crohn’s disease as the main reason of failure of this operation, which was diagnosed in 15% of the patients with pouchitis and pouch failure. He also proved that indeterminate colitis and intraoperatively diagnosed terminal ileitis are not always associated with CD and failure of restorative proctocolectomy (18). Nagar also advocates the need for earlier surgical intervention for children suffering from UC (17). The operation results in a fast withdrawal of the symptoms, allows for the patient to resume school activities, improves quality of life and facilitates the termination of aggressive forms of pharmacotherapy (steroids and immunosuppressants) (7). Nagar stresses the low complication rate and very good functional results includ-
ing intact sphincter function and less than 4 stools per day

CONCLUSIONS
1. Surgical treatment of IBD in children has become an accepted method of treatment, and is sometimes the only treatment which can cure the severe complications of these diseases.

2. Restorative proctocolectomy with IPAA is the treatment of choice in adults with UC. This operation should also be considered more often in children with UC prior to conservative treatment, especially since steroids and immunosuppressants produce undesired side-effects

3. A consulting surgeon should be involved in the treatment of IBD in paediatric patients at an earlier stage than is currently practiced.

REFERENCES


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COMMENTARY

Both Crohn disease and ulcerative colitis are the inflammatory bowel diseases. To the causes of the both diseases are proposed: genetic, environmental, immunologic as well as infectious etiologies.

Inflammatory changes in ulcerative colitis are only in large bowel mucosa. These changes in Crohn disease are in the full thickness enteric wall and can involve every part of the alimentary tract, but its main location is in the ileo-cecal portion. It is believed that the disease onset in 15-20% of patients is in the childhood. The incidence of the inflammatory bowel disease is increasing. The section of inflammatory
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bowel disease has been founded in the Polish Association of Pediatric Gastroenterology, Hepatology and Nutrition. The diagnostic and therapeutic criteria including indications to the surgery of the inflammatory bowel disease have been established in the Section forum. These indications depend mainly on the severity and duration of the disease. They are significantly different for both entities.

The index assessing child’s nutrition status and Pediatric Crohn Disease Activity Index (PCDAI) are used for the evaluation of the severity of the disease. PCDAI assess clinical symptoms, inflammatory parameters and child’s nutrition degree. The basic management is constantly modified pharmacological therapy. There is however believed that ca. 50% of children need surgical treatment in 3-5 years after the onset of the disease. The indications to the surgery can be divided into two groups. The first group are emergencies like ileus, perforation, intraabdominal abscess, and rarely enteral bleeding; the second group are elective procedures: no relief after conservative treatment of the acute disease crisis, intestinal stenoses and fistulas. Discussing elective indications one should remember that surgical treatment does not guarantee complete recovery and most patients will need next operations leading to the short bowel syndrome. The surgical treatment does not discharge the need of the simultaneous conservative treatment. Resections and anastomoses should be performed based on surgical principles; in the case of Bauhin valve resection, isoperistaltic small bowel valve should be created. Special surgical problem is the first recognition of the Crohn disease during the operation for the suspected appendicitis. The management depends on the subjective assessment and experience of the surgeon and on the general condition of the child. It seems that ileocecal resection is justified when inflammatory changes of the cecum are obvious. Appendectomy as the only procedure threatens the production of the intestinal fistula or abscess and the disease recurrence after ileocecal resection is observed later after longer duration of the remission.

Ulcerative colitis is assessed by Truelove and Witts scale complying clinical presentation, inflammatory parameters and ischemia degree. The disease is classified as mild, moderate and severe forms. Pharmacologic treatment gives improvement in ca. 80% cases. The indication for emergency, life-preserving operation is acute sepsis with paralytic ileus called as toxic megacolon. The emergency indication for the operation can be also the perforation and rarely stenosis and fistula. However, often recurrences leading to the growth failure and the delayed maturation are the elective indications to the surgery. For the favor of the surgery is the fact that surgical intervention in the ulcerative colitis, as opposed to the Crohn disease is curative and further conservative treatment is unnecessary. One should also remember that after 10 year of the disease the risk of neoplastic metaplasia is increasing significantly. The surgical intervention is complete colectomy. We performed it based on the Soave procedure described for the Hirschsprung disease. The whole colon to the level of the rectum is resected from the abdominal cavity aspect and the rest of the mucosa is removed from the perineal aspect. The small bowel is pulled through the created channel. In this way both internal and external sphincters are intact. We create J pouch before the small bowel anastomosis, Kaliciński-CZD – does not create the pouch but creates invagination valve. The surgical results are good as far as the stool frequency and continence are concern. When the operation is elective and performed after good preparation of the child, even temporary ileostomy is not necessary.

The child with inflammatory bowel disease needs also nutritional management and permanent psychological care besides pharmacological therapy both before and after operation. Therefore such a child should be treated by the experienced team lead by pediatric gastroenterologist with active presence of pediatric surgeon. This model is realized in large pediatric centers in our country. Therefore, appreciating the author’s center, where “generally” and “everyday” adult patients are treated, I would like to stress finally that children with inflammatory bowel disease should be referred to the tertiary pediatric centers.

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