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Lecture

Inflammatory Bowel Disease in Childhood: Aspects of Surgical Therapy

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SUMMARY

Following the diagnosis as exactly as permitted, many children will have to undergo corrective surgery, mainly dependent on the complications of CD or UC respectively and/or the intractability to conservative therapy. Surgery may be corrective mainly in CD cases or colectomy if the disease is restricted to the large bowel for CD. In cases of UC colectomy is curative. The indication and results of surgical intervention is discussed.

Key words: Inflammatory Bowel Disease, Children, Surgery

It is almost 200 years since the first report of a disease, which some 100 years later was described as regional enteritis and was given the name of its first author: Crohn's Disease (CD). The history of Ulcerative Colitis (UC) is almost as long. It was initially thought to be completely distinct from CD. Gradually however, common features were recognized, in addition to those separating the two entities. Also, there remain some cases where the distinction between the two is impossible or remains so for a considerable time, delaying the final diagnosis. So now they go together under the term Inflammatory Bowel Disease. Based on these facts CD and UC are usually considered together.

The features (common and distinct) of CD and UC have been sufficiently described and are therefore presented only in summary (Table 1). The etiology for both has not yet been clarified. It involves in both diseases an inflammatory process, which allows grouping them together under the same heading. The pathology has also been described; suffice it to say in broad terms, that the inflammatory process in CD involves the entire thickness of the intestinal wall, while UC affects primarily only the mucosa. In addition, CD may affect any and all segments of the intestinal tract, which explains the term regional enteritis, whereas UC is always restricted in the large bowel leaving the proximal parts of the intestinal tract free.

Differences between the two entities determine to a great extent the planning of the treatment of each of these two diseases, particularly with respect to surgery. Crohn's Disease is characterized by its regional involvement. Its manifestations can usually be quite successfully treated by non-surgical means, however, a significant percentage of these patients will need surgery some time during the course of their illness. The scope of pharmacologic treatment and its results remains in the hands of the pediatrician-gastroenterologist. However, quite characteristic of CD is the frequent appearance of complications, which are the main determinants for surgery.

Prediction of those patients who may develop complications and need subsequent surgery is difficult. Many factors have been investigated as to the predictability of early surgery as well as the possibility for recurrence requiring surgery. Genetic factors, such as CARD15 and other mutations have been positively connected with early surgery,[1] as has been antisacharomyces cerevisiae antibody (ASCA).[2] Duration of the disease positively increases the risk for surgery.[3] Other factors which seem to increase the risk for surgery include female gender, an initial diagnosis as UC, poor growth at presentation. On the other hand small age, preoperative treatment with infliximab and/or 5-aminosalicylic

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Table 1. Main pathologic differences between Crohn's Disease and Ulcerative Colitis

	Crohn's Disease	**Ulcerative Colitis**
Etiology	**Unknown**	**Unknown**
Involvement	Regional involvement of any GI segment (*In 10-15% localized in the colon*)	Continuous involvement of the large bowel (*The terminal ileum may be affected by refluxing material*)
Extra intestinal manifestations	*Relatively common:*	*Relatively uncommon:*
	Weight loss	Weight loss
	Growth retardation	Growth retardation
	Delayed puberty	Delayed puberty
	Hepatitis	Hepatitis
	Arthritis	Arthritis
	Relatively uncommon:	*Relatively common:*
	Sclerosing Cholangitis	Sclerosing Cholangitis
Complications	*Relatively common:*	*Relatively uncommon:*
	• Adhesions betw. Intestinal loops	• Adhesions betw. Intestinal loops
	• Abscess formation	• Abscess formation
	• Strictures	• Strictures
	• Fistulae	• Fistulae
	Relatively uncommon:	*Relatively common:*
	• Toxic megacolon	• Toxic megacolon
	• Dysplasia	• Dysplasia
	• malignancy	• malignancy

MATERIAL

J pouch	2
S-pouch	1
Straight ileo-anal anastomosis (non IBD included)	6 (2)
Crohn's colectomy	2
Crohn's enterectomy	1
Hippocratic fistula ligation	1
Total cases	12

acid seem to have a positive predictive value.[4,12] Restriction of CD in the colon may also be a negative prognostic factor for surgery.[5]

Fistula formation, either external or internal, is a quite frequent complication of CD. It is the sequel of the transmural involvement of the bowel. These fistulas may be resistant to non-surgical treatment and may consequently require surgical excision. This excision may involve enterectomy, which can be rather extensive and anastomosis. It is therefore mandatory to try to bring the basic disease as much as possible under control before attempting resection. External fistulae appear usually in the form of perianal fistulae. Ligation and traction by means of the Hippocratic method may be beneficial; otherwise, deviation of the fecal stream by means of an enterostomy may become necessary. Abscess formation, another frequent complication of CD, may in its initial stages respond favorably to non-surgical treatment, but will often require drainage and if it results in the formation of a fistula then resection will be the treatment of choice.

The planning of such operations may be quite difficult and considerations involved include strictureplasty and/or resection and anastomosis. A significant factor influencing the decision is postoperative prognosis and surgical effectiveness in a disease which is chronic and relapsing.[6] Surgical correction of complications and intractable disease seems to have a positive effect on growth and puberty, which may previously be retarded, although this may not reach statistical significance.[7]

Stricture formation is another frequent complication of CD leading to incomplete or even complete intestinal obstruction. Depending on the extent and severity of the stricture one may choose between strictureplasty or resection and anastomosis. At times even the placement of a temporary enterostomy may appear advisable. There is some controversy concerning the extent of resection, there is an increased tendency towards spearing resection, leaving macroscopically less involved margins. Studies involving short term compli-

cations and long term outcome and the need for reoperation indicate that there is no significant difference in short term complications between radical and bowel conserving resection.[8] In addition, it does not influence the need for subsequent operations. In one study there seems to be a difference depending on the type of anastomosis, the end-to-side and side-to-side anastomosis leading to better results,[9] but this is not supported by other studies.

A significant prognostic factor as to the risk for an initial operation is age as well as the duration and severity of the disease.[3,8,10] Accordingly, children have a favorable prognosis as to the need for surgery.[7,10] This explains a relative paucity of literature concerning pediatric patients alone. It also explains the relatively small number of children treated surgically in our institution.

The outcome of these operations is favorable and may result in significant improvement of growth as well as development of puberty.[7,19] However, partly due to the chronic relapsing course of the disease, there is a high risk for recurrence and hence the need for reoperation. Many factors have been investigated regarding this chance. The existence of perforation at the time of initial surgery was an unfavorable predictive sign for the need of subsequent operations.[11,12] Other unfavorable predictors include ASCA,[13,2] duration of the disease, existence of perforation at initial surgery,[22] duodenal and jejunal involvement.[8] In one study involving children, Pediatric Crohn's Disease Activity Index at the time of surgery and the use were 6-Mercaptopurine (6-MP) of independently associated with higher recurrence rates.[14]

In general terms, surgical treatment of the complications of CD will require careful planning and close collaboration between the pediatrician/gastroenterologist and the surgeon for optimal results. One must bare in mind that "regional enteritis" may present with relapsing episodes interspersed with periods of remission and that normal appearing segments of the intestine may either flair up in the future or become involved requiring further resection. It is therefore advisable during surgery to be as conservative as possible.

By contrast, Ulcerative Colitis is characterized by strict involvement of the large bowel. Therefore, its elimination will result in permanent cure of the disease. In addition, it is well established that in long standing active disease the development of colonic cancer is a frequent complication. The cumulative risk for the development of carcinoma in pediatric patients has been estimated to increase by about 10% with every decade of active disease.[17,15,16] In one recent publication, this risk is reported to be significantly less.[18] Nevertheless, the standard treatment of UC, as in CD, remains pharmacologic and surgery is indicated either in long standing active, intractable disease, or in the presence of complications. In such cases resection of the diseased colon is recommended.

Toxic megacolon is the most serious complication. It is reported to occur in about 5% of the cases and is even more infrequent in children. However, it is a medical and surgical emergency and needs to be treated aggressively. It is characterized by colonic dilatation, accompanied with general toxicity, fever and abdominal tenderness as in peritonitis. Due to the toxicity, cardiac involvement, electrolyte disturbances and dehydration may confound the problem. The greater the colonic dilatation, the greater the risk of perforation, which is accompanied by increased mortality. The patient's homeostasis must be restored as quickly and as well as possible, together with bowel rest and parenteral nutrition. If treatment fails to improve the patient's condition, or if perforation or hemorrhage supervenes, emergency colectomy will be required. Otherwise interval colectomy will have to be considered.

Another indication for colectomy is intractable disease expressed as chronic dependence of steroids. Lastly, in children, growth factors and the retardation of puberty must also be included in the equation. The benefits of total colectomy and the improvement of life substantially outweigh the risks of surgical colectomy. In most children with growth and puberty retardation the development is favorably restored following resection.[7,19] In addition, colectomy represents the only cure of the disease and thus fully justifies the surgical intervention.

The operation consists of total colectomy and ileoanal pull through with rectal anastomosis. Ideally it is performed in two stages. In the first stage colectomy and terminal ileostomy is performed. The rectal stump is exteriorized suprapubically as a mucus fistula. This results in dramatic improvement of the patient's general condition. When the mucosa in the rectal stump has sufficiently improved by endoscopy, then the second stage may be achieved, consisting of stripping off the mucosa of the stump and pull through of the terminal ileum through the muscular cuff. The ileo-rectal anastomosis is performed right above the dentate line of the rectum.

The creation of a reservoir in terms of a J-pouch, S-pouch or W-pouch remains controversial. Most authors

prefer the creation of a J-pouch,[20,21] which is simpler and carries less complications than the other more elaborate techniques while being functionally quite comparable. In our impression, there is no need to create a reservoir for continence because the terminal ileum responds well in its function of fluid reabsorption and after a relatively short period of time bowel movements are reduced to <5 per day with interval continence. In the small number of 12 patients operated upon in our department, we encountered 2 cases of pouchitis (Table 2). Both children turned out to have CD. One responded to non-surgical measures, while the other had to be converted to an ileostomy. By contrast, in 6 children submitted to straight ileoanal anastomosis (including 2 with familial polyposis), there was no pouchitis or other complication. All children were continent with 3-5 stools daily, after a short period of adjustment. Care must be taken to keep the muscular cuff short in order to avoid stenosis at the level of the pulled through segment of the ileum.

The prognosis is good in most cases of both CD and UC. Patients with CD may expect to live a normally long life, although morbidity may lead to frequent hospitalizations. This is particularly true in predominantly colonic involvement. In UC many patients lead a normal life with some pharmacologic support. In severe or long standing cases total colectomy will be curative. After a period of adaptation, the increased frequency of stools will be well tolerated.

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