



HIRSCHSPRUNG'S DISEASE; ENTEROCOLITIS DURING THE COURSE

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ABSTRACT... Objective: To determine the frequency of Hirschsprung's Disease associated enterocolitis in pre and post operative patients. **Place and Duration of Study:** Nishtar Hospital and Ibn-e-Sina Hospital, Multan from January 2008 to June 2012. **Methodology:** A total number of 114 patients aged from less than 6 months to more than 5 years were diagnosed and treated for Hirschsprung's Disease after rectal biopsy and internal sphincterotomy. **Results:** In total of 114 patients 32 (28%) developed enterocolitis before and 5 (18.5%) after definite surgery. **Conclusions:** The results of this study implicates that the early diagnosis and to make measures to decrease the post pull-through anastomotic anorectal stenosis can reduce the frequency of pre and post operative Hirschsprung's disease associated enterocolitis.

Keywords: Hirschsprung's Disease, pre and post operative enterocolitis.

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INTRODUCTION

Hirschsprung's disease associated enterocolitis remains a major source of morbidity and mortality both before and after definite surgical treatment¹. A lot of work has been done on the different aspects of pre and post surgical Hirschsprung's disease associated enterocolitis^{2,3}.

In this study we have endeavored to see the frequency of enterocolitis in pre and post surgical cases where the diagnosis was late or anorectal stenosis did develop after endorectal pull-through leading to enterocolitis.

MATERIAL AND METHODOLOGY

A total number of 114 cases i.e. 90 male and 24 female patients were admitted and treated in the department of paediatric surgery, Nishtar Hospital and Ibn-e-Sina Hospital, Multan. The age at the time of diagnosis is given in table-I. The diagnosis was established by rectal full thickness biopsy. In this study the colostomy was always performed in the ganglionic segment, as there were no means available to confirm the level of aganglionosis. The dilated portion was considered to be the safe side

for colostomy. In this study, 63 underwent for laparotomy and the remaining patients were either of ultra short segment or having no Hirschsprung's Disease. In majority of cases (32), transverse colostomy was done. In three cases of total colonic aganglionosis, ileostomy was performed and in the remaining cases the colostomy was performed accordingly. Soave's pull-through was performed in 27 patients while 34 are waiting for surgery.

Age	No. of patients
Less than months	38
6 to 24 months	21
2 to 5 years	49
Above 5 years	06

Table-I. Age at the time of diagnosis

RESULTS

27 patients had the history of enterocolitis before surgery. No other obvious cause was detected except the late diagnosis. 63 cases underwent for laparotomy and colostomy/ileostomy. The extent

of aganglionic lesion is shown in table 2. Among 27 patients who underwent for definite pull-through operations, 5 developed anorectal stenosis and these patients were also manifested with enterocolitis.

Rectum	26 (41.2%)
Rectosigmoid	23 (36.5%)
Pelvic colon	7 (11.11%)
Transverse colon	2 (3.1%)
Total colon	3 (4.7%)

Table-II. Extent of Aganglionic Lesion

DISCUSSION

Now Hirschsprung's Disease associated enterocolitis has been recognized as the well known complication of Hirschsprung's disease before and after surgery. Usually it starts in infancy⁴ but it can occur in any age. Enterocolitis with Hirschsprung's Disease is characterized by watery diarrhea with the passage of yellow foul smelling stools. The patient fails to gain weight and become lethargic. The onset is heralded by fever, abdominal distension and prostration due to acute water and electrolyte imbalance⁵. It is believed that enterocolitis is more common in long segment Hirschsprung's disease, but Teitelbaum et-al⁶ did not find any relation to the level of aganglionosis. Similarly Carneiro et-al⁷ also noted that long segment aganglionosis was not associated with an increased risk of enterocolitis. However the association of trisomy is well recognised⁸.

Significant advances in the diagnosis and management of Hirschsprung's Disease have been laid. Enterocolitis which may be pre or post operative is still a major cause of morbidity and mortality⁹. The pathology of Hirschsprung's Disease associated enterocolitis is still poorly understood. Where various hypotheses i.e. hypersensitivity reaction to bacterial antigen, proximal dilatation with mucosal ischaemia and bacterial invasion, enterocolitis and prostaglandins, defective immunological defense, impaired gastrointestinal mucosal defence and alterations in intestinal mucins have been

advocated¹⁰.

In this study it was considered that proximal dilatation with bacterial overgrowth is the leading factor in the development of pre and post operative cases. Hirschsprung's Disease associated enterocolitis is commonly manifested in delayed diagnosed cases and also in post pull-through cases where stenosis develops after anorectal anastomosis¹¹. If diagnosis is delayed and primary pull-through or diverting colostomy is not performed early there will be continuous dilatation of proximal gut followed by the bacterial overgrowth. Same pathology develops when anorectal stenosis is developed at the level of anorectal anastomotic site. It is therefore considered that this post pull-through complication is the causative factor of Hirschsprung's Disease associated enterocolitis¹²⁻¹³. In our study 37 patients developed enterocolitis, 32 (28%) in pre and 5 (18.5%) in post operative cases. In some reports¹⁴ the incidence is 25%, reported greater incidence than 70% to 50%¹⁵. Arnold et-al¹⁶ reported 17-18% enterocolitis in post operative cases. Over the past 40 years there has been clear decline in the incidence of Hirschsprung's Disease associated Enterocolitis due to improved techniques, regular follow-up and early diagnosis. We have also analyzed the reasons behind the late diagnosis and development of anorectal stenosis.

In our circumstances the patients became late, perhaps many patients in our circumstances are treated by the general practitioners and paediatricians at various levels either as diarrhea or chronic constipation. Diarrhea is still one of the common problem in this country and carries the high mortality. Some of the patients with Hirschsprung's Disease associated Enterocolitis are not reported to paediatric surgical units. Similarly patients with chronic constipation also do not report and treated with laxatives and enemas etc.

Another factor for poor referral is non-availability of qualified doctors in rural areas. In this setup described, the delayed diagnosis is obvious. Anastomotic stricture is a late major complication

of pull-through operation¹⁷. This mechanical factor becomes the main risk factor in the development of post operative Hirschsprung's Disease associated Enterocolitis. This happens due to surgical error or to the poor compliance of post operative dilatation. Parents were advised to dilate the anal canal with dilators at home, until the anocolic anostomosis remained at a satisfactory caliber. The parents were advised to report for follow-up. Failure to follow the instructions resulted in above mentioned complication.

Hirschsprung's Disease associated Enterocolitis is frequently treated with rectal wash outs, administration of broad spectrum antibiotics and regular anal dilatations. In failure of these measures, internal sphincterotomy has to be performed¹⁸. Realizing this we performed the internal sphincterotomy jointly with rectal biopsy to overcome the future internal sphincter spasm, which is a major factor of post-operative enterocolitis.

This study reflects that among the 27 of the cases presented with late diagnosis (Table 1) developed enterocolitis. Ideally all these cases should have been diagnosed in neonatal age prior to the development of stasis, bacterial overgrowth and invasion of gut wall. In all these cases colostomy/ileostomy should have also been established in neonatal age to avoid the future consequences of enterocolitis. In present study 5 patients developed anorectal stenosis followed by enterocolitis after 27 pull-through operations. Surgery is a personal maneuver with varying results of individuals. All the surgical techniques are good in the hands of experts. If the principles of surgery are followed accordingly, then there is no question to have decrease in post-operative complications. Last but not the least, parents should be educated and be realized about the importance of regular follow up.

CONCLUSIONS

It has been emphasized that the pre and post-operative enterocolitis develops due to intestinal

stasis with the proliferation of luminal microbes and ultimately invasion of intestinal mucosa. Intestinal stasis may be due to post pull-through anorectal stenosis or due to delayed diagnosis, where the gut dilates above the aganglionic contracted segment resulting partial obstruction and the final outcome as enterocolitis. So the frequency of pre and post enterocolitis is related to diagnostic delay and the consequences of technical error i.e. post pull-through anorectal stenosis.

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It is better to fail in
originality than to
succeed in imitation.

Herman Melville

