

**LONG TERM FOLLOW-UP OF HIRSCHSPRUNG'S DISEASE:
REVIEW OF EARLY AND LATE COMPLICATIONS**

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Objective: Long term follow-up of Hirschsprung's disease to determine the early and late complications. **Design:** Retrospective. **Setting:** Tertiary care referral hospital. **Subjects:** Fifty children who underwent either Swenson's procedure [SP] (n=16) or Martin's modification of Duhamel's procedure [MDP] (n=34), for treatment of Hirschsprung's disease during the period 1981 through 1990. **Results:** Early complications observed were anastomotic leak (n=4), stenosis (n=8), wound infection (n=8), and intestinal obstruction (n=6). Follow-up period ranged from 2-10 years. Long term complications included constipation (n=1), enterocolitis (n=2), mild soiling (n=4), and intestinal obstruction (n=2). **Conclusions:** Early complications were seen with equal frequency in patients with either SP or MDP but late complications were seen less often with the SP. Both SP and MDP were equally effective in the definitive treatment of Hirschsprung's disease.

Key words: Hirschsprung's disease, Swenson's procedure, Duhamel's procedure, Complications.

DESPITE advances in surgery, anesthesia and post-operative management, surgical treatment of Hirschsprung's disease may result in some early and late complications in the form of anastomotic leak, stenosis, abdominal wound infection, constipation, enterocolitis and death. Since the introduction of the pull-through operation by Swenson(1), there have been a number of papers on Hirschsprung's disease highlighting the complications of various procedures. There has been no large long-term series from India. The experience with Swenson's procedure [SP] and Martin's modification of Duhamel's procedure [MDP] in 50 patients over the past ten years, from 1981 through 1990, in the Department of Pediatric Surgery, All India Institute of Medical Sciences [AIIMS], New Delhi,

has been presented here.

Subjects and Methods

The hospital records of 50 patients of Hirschsprung's disease, treated with SP or MDP, were reviewed for early and late complications. All the SP were performed by one surgeon (VB) and all the MDP were performed by another (DKM), in the same period of time, depending on whose out-patient the case presented to. Both the groups were well matched. All these patients were called for follow-up. Twenty six of the 50 reported for follow-up. Of the remaining 24, one child a follow-up of SP had expired and the other 23 responded but informed of their inability to come and provided details of their latest clinical status.

Early complications were defined as those which occurred within 30 days of surgery. Anastomotic stenosis was defined as any narrowing of the anastomosis that required dilatation or revision. Sudden onset of diarrhea, abdominal distension, fever and leukocytosis was considered to be enterocolitis. Abdominal distension associated with constipation, vomiting and increased bowel sounds was labeled as intestinal obstruction. Involuntary passage of fecal matter with or without gas was considered as soiling.

Results

There were a total of 50 patients—44 (88%) males and 6 (12%) females. The level of aganglionosis extended proximally up to the sigmoid in 26%, recto-sigmoid in 50% and rectum in 10% patients. In the remaining 14% this level varied from the descending to the transverse colon. A preliminary diverting colostomy was performed in all patients. The definitive treatment was by SP in 16 patients and MDP in the other 34 cases.

The early complications observed are shown in *Table I*. There was no mortality. Anastomotic disruption was seen in 2 of SP and 2 of MDP. All these 4 patients had undergone a previously failed myectomy for treatment of Hirschsprung's disease. Anastomotic stenosis occurred in the 2 patients of SP procedure who developed a leak. Five of the 34 patients of MDP also developed mild stenosis but none of these had an obvious anastomotic leak. Early intestinal obstruction was seen in 2 of SP and 4 of MDP patients. All the 6 patients improved on conservative management.

All the 50 patients were evaluated for long-term results. Of these, 26 patients who came for follow-up were clinically assessed, while in the remaining 23 patients the evaluation was based on a simple questionnaire. One patient, a follow-up of SP had expired in 1990, seven years after pull-through, in a district hospital where he was admitted with adhesive intestinal obstruction. Of the 26 who reported, 7 were of SP and 19 of MDP. Six of the 7 (85.7%) of SP and 13 of 19 (68.4%) of MDP

TABLE I—Complications Following SP and MDP

Complication	Procedure		
	SP (n=16)	MDP (n=34)	Total (n=50)
<i>Early</i>			
Anastomotic disruption	2 (12.5)	2 (5.9)	4 (8.0)
Stenosis	2 (12.5)	6* (17.7)	8 (16.0)
Abdominal wound infection	3 (18.8)	5 (14.5)	8 (16.0)
Intestinal obstruction	2 (12.5)	4 (11.7)	6 (12.0)
<i>Late</i>			
Constipation	0 (0.0)	1 (3.0)	1 (2.0)
Enterocolitis	0 (0.0)	2 (5.9)	2 (4.0)
Incontinence (fecal)*	0 (0.0)	4 (11.4)	4 (8.0)
Intestinal obstruction	1 (6.3)	1 (3.0)	2 (4.0)
Mortality	1 (6.3)	0 (0.0)	1 (2.0)

Figures in parentheses indicate percentages.

* One patient developed ischemic narrowing of the pulled through colon.

+ There was no case of urinary incontinence with both procedures.

defecate once a day. One follow-up patient of SP and 4 of MDP defecate 3-4 times a day. All these children are less than 5 years of age and less than 24 months post-operative. One child, a follow-up of MDP still has a colostomy and is awaiting re-pull-through as his previously pulled-through colon is very narrow and atrophic, possibly, secondary to ischemic damage. Another patient a follow-up of MDP had persistent constipation due to stenosis and required regular enemas and dilatations. The 23 children who responded to the questionnaire but did not come for follow-up have normal bowel habits.

No patient in either of the groups has urinary incontinence, although mild fecal soiling is present in 4 of the MDP patients. One episode of post-operative enterocolitis was observed in each of two children following MDP and both improved on conservative management. One patient in each of the groups developed late intestinal obstruction due to adhesions; the one with SP having expired as mentioned earlier.

Discussion

Prevention of constipation with preservation of sphincter function is the therapeutic aim in Hirschsprung's disease. The definitive treatment is by either primary pull-through or by a preliminary diverting colostomy followed by pull-through. At AIIMS the latter approach has been followed as most of our patients present with long standing obstruction, moderate to severe malnutrition, severe colonic hypertrophy and dilatation and the colostomy helps in the recovery of these children. It also helps in clearing the distal colon of fecalomas and thereby the colonic hypertrophy tends to resolve, making the colon more amenable to pull-through.

The physiologic principles embodied in

the Swenson's operation(1) have been corroborated many times. Refinement in the technique and the addition of partial internal anal sphincterectomy(2,3) to the original procedure has improved overall results. The original Duhamel's procedure(4) implied complete division of the internal anal sphincter, but this caused a high incidence of incontinence. This prompted a series of modifications to partially preserve the internal anal sphincter(5-8). The Martin's modification(8) was introduced to take care of the blind rectal pouch and the problems associated with it. Martin's modification of Duhamel's procedure was used in our patients.

The incidence of anastomotic leaks has been reported to be 5-15% following SP(9-13) and 2-10% following MDP(9-14) in various series. In the present series it was 12.5% for SP and 5.9% for MDP. All our patients who developed a leak had a previous myectomy which might have caused increased scarring and poor healing at the anastomotic site. The reported incidence of stenosis is 6-15% following SP(9-11,13,15) compared to 12.5% in the present study. The incidence of stenosis reported for MDP is 5-15%(9-11,13) as compared to 14.7% in the present report. The incidence of abdominal wound infection in this series is higher than that reported in the literature(10-14). This may be because of the poor nutritional status and presence of a colostomy in all our patients.

The incidence of early obstruction was similar in both the groups in this study (12.5% following SP and 11.7% following MDP) as compared to the reported incidence of 3-12% for SP(10,12,13,16) and 7-20% for MDP(10,12,13,14). All our patients responded to conservative management.

The incidence of persisting post-operative constipation has been reported as 6-15% in SP(4,11,13,15,17,18) and 4-10% in MDP(4,11,13,15,18).

There are three main factors in the causation of post-operative constipation; (i) Until the sensation of feces in the neorectum is appreciated by the child he will ignore it and, therefore, fail to empty the terminal bowel. To avoid this, the patient must undergo intensive toilet training in the initial few post-operative weeks; (ii) The colon proximal to the point of resection may be considerably dilated and likely to empty incompletely due to colonic inertia. Diverting colostomy as a first stage procedure is particularly helpful in this regard. The decompressed colon returns to normal caliber and function; and (iii) Distal resection should include partial sphincterectomy to take care of the internal anal achalasia.

Soiling in the initial post-operative months is a common problem as most patients following resection tend to have large volume of liquid intestinal contents. Passage of flatus in this situation invariably causes some soiling. Gradually the intestinal contents become more solid and soiling is eventually eliminated. In young children who are not toilet trained, the observation that the child is clean between bowel movements is taken to indicate that sphincteric control is present. Four of our patients of MDP, have mild persistent soiling which is gradually improving. Total fecal incontinence is usually because of sphincter damage or spurious diarrhea associated with fecal impaction. Attention to surgical detail and avoiding impaction will take care of this problem. The reported incidence of fecal incontinence is 2-20% for SP (9,11-13,15,17,18) and 1-14% for MDP (9,11,13,15,17). Late intestinal obstruction is usually because of adhesions and unrecognized strangulation is the cause of many a late deaths. It was the cause of the only mortality in this series.

In conclusion, both SP and MDP are equally effective in curing Hirschsprung's disease and both procedures have a similar incidence of complications.

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