Rehbein's Procedure in 73 Cases of Hirschsprung's Disease

Somchai Thepcharoennirund MD, FRCST*

** Pediatric Surgical Unit, Department Of Surgery, Ratchaburi Hospital*

The principle for treatment of Hirschsprung's disease by Rehbein's procedure is to remove the aganglionic narrow segment including the dilated sigmoid colon by dissection of the upper rectum deep down into the pelvic cavity about 2 cm from peritoneal reflection and to elimination of the achalasia of internal anal sphincter by vigorous dilatation. Between August 1987 and April 2000, 73 cases of Hirschsprung's disease (50 boys and 23 girls) were operated on by Rehbein's procedure at the Pediatric Surgical Unit, Ratchaburi Hospital. The age varied from 7 months to 10 years. 55 cases showed very good results by daily stool evacuation without any help, occasional laxative or enema in 15 cases, giving laxative or enema regularly in 2 cases and no adequate bowel evacuation despite giving laxative or enema in 1 case. In conclusion, Rehbein's procedure is technically simple and has good results. It should be another treatment of choice for Hirschsprung's disease.

Keywords : *Rehbein's procedure, Hirschsprung's disease*

J Med Assoc Thai 2004; 87(10): 1188-92
e-Journal: *http://www.medassocthai.org/journal*

Hirschsprung's disease is a congenital anomaly of the colon affecting about 1:5,000 live-births[1,2]. Harald Hirschsprung in 1886 gave the first detailed description of congenital megacolon. The term "megacolon" was introduced by Mya in 1894. In 1901, Tittel described the absence of intramural ganglion cells in the rectum. Ehrenpreis' 1946 doctoral thesis, an exhaustive review of the etiology and pathogenesis of Hirschsprung's disease, pointed out that the diagnosis could be made in the neonate[3,4,5]. The aganglionosis leads to sustained contraction of the diseased colon with secondary dilatation and hypertrophy of the more proximal segment. "Congenital megacolon" was named after this secondary change[6]. In 1948, Swenson and Bill presented a curative operative technique. Professor Fritz Rehbein developed a procedure for Hirschsprung's disease that bears his name in 1953. During his career he successfully used this operation on nearly 400 children. His procedure remains the operation of choice in many pediatric surgical centers world wide[3]. Duhamel in 1956 proposed partially bypassing the rectum and performing the anastomosis with the use of crushing clamps. Soave in 1962 applied the endorectal pull-through procedure for Hirschsprung's disease[7,8,9,10]. Textbooks of pediatric surgery never mention Rehbein's procedure. But Swenson, Duhamel, Soave and Rehbein have published their present views on the surgical procedures they introduced in the book "Hirschsprung's Disease", of which Professor Alexander Holschneider was the editor[4].

The principle for treatment of Hirschsprung's disease by Rehbein's procedure is to remove the aganglionic narrow segment including the dilated sigmoid and to eliminate the achalasia of the internal anal sphincter[4]. The purpose of the present study was to re-emphasize the author's modification of the Rehbein's procedure and report the result of treatment with Rehbein's procedure in Ratchaburi Hospital.

Material and Method

Patients and procedure

Between August 1987 and April 2000, 73 cases of Hirschsprung's were operated on by Rehbein's procedure at the Pediatric Surgical Unit, Department of Surgery, Ratchaburi Hospital. The author does not routinely perform colostomy in every case. It must be done in severe ileus and when enterocolitis occurs. Colostomies were necessary in 22 patients in the newborn period (16 sigmoid colostomies and 6 descend-

Correspondence to : Thepcharoennirund S, ***Pediatric Surgical Unit, Department Of Surgery, Ratchaburi Hospital, Ratchaburi 70000, Thailand. Phone: 0-3232-2937, 0-4010-2979, Fax: 0-3232-1825***

ing colostomies) and ileostomies were done in 2 patients due to total colonic aganglionosis. Details of operative technique and long term follow-up have been tabulated.

Operative technique

Rehbein's procedure (Modification in Ratchaburi Hospital)

1. Preoperative Sphincter Dilation

Vigorous sphincter dilation is performed on the operating table immediately prior to surgery using a spreading instrument or, in older children, digitally under general anesthesia. Placement of an indwelling urinary catheter is necessary before surgery.

2. Positioning

A sand bag or folded towel is placed directly under the buttocks. The thighs are fixed loosely to the operating table by adhesive strips above the knees.

3. Opening the abdomen

The abdominal cavity is opened using a low transverse incision.

4. Preparation of Surgical Field

The lateral umbilical ligament is divided on both sides after ligation. An abdominal wall retracter is introduced.

5. Dissection of the Upper Rectum

The peritoneum is divided on the left and right sides of the upper rectum, followed by the peritoneum reflection. The superior hemorrhoidal vessels are ligated and cut. The distal limb of the sigmoid is pulled up vigorously in order to obtain a good view of the upper rectum. The perirectal tissue is separated until the external longitudinal muscle layer of the rectum is exposed. The dissection is completed when 2 cm of the upper rectum in infants and 3 cm in older children, measured from the peritoneal reflection, are freed up (Fig. 1).

6. Dividing the rectum

The rectum is stretched vigorously by tension on the sigmoid, and initially divided transversely to the midline, starting from the right side. Holding sutures are placed at the lower border of the incision on the right, anteriorly and posteriorly. Following complete division of the rectum, another holding suture is placed on the left incisional border.

7. Dividing the Descending Colon

The large bowel is usually divided in the lower portion of the descending colon including sigmoid colostomy (Fig. 2). The descending mesocolon is cut up to the left colic artery and vein. A strong clamp is placed just above the sigmoid, and the descending colon is divided. Four holding sutures are also placed here, at the upper border of the descending colon incision. The large bowel is cleaned using several moist sponges.

8. Colorectal Anastomosis

The descending colon is brought down to the severed rectum with the aid of the 4 holding sutures, and the anastomosis is performed using continuous 4-0 vicryl sutures interrupted at 4 sites. The four interruptions of the sutures are expedient in preventing stenosis of the rectum at the suture area.

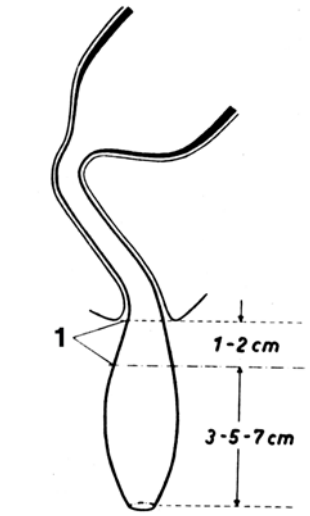

Fig. 1 A segment 1 to 2 cm long is dissected free below the peritoneal reflection (1). The distance of the cut level from the anal orifice varies according to the child's age. It is about 3 cm in infants and somewhat greater in older children

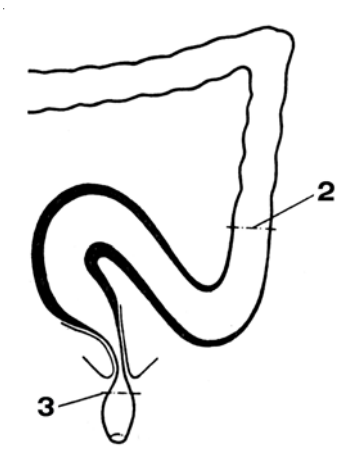

Fig. 2 Extent of resection. The large bowel is divided proximally at the distal descending colon (2). (3) Level of division of the rectum

The right half of the posterior wall is sutured together, followed by the left half (Fig. 3,4). A soft synthetic tube about the size of the little finger is now inserted down the still-open anastomosis, through the rectal stump and out the anus until the tip of the tube lies at the level of the anastomosis. The tip of the tube is then pushed 5 to 8 cm upward into the descending colon, again via the open anastomosis. The two halves of the anterior wall are each joined using continuous sutures (Fig. 5,6). There is no difficulty in performing the anastomosis when the right and left corners of the rectal stump are pulled up during suturing. After the sutures are cut, the anastomosis moves back anally and extraperitoneally. At the end of surgery, the rectal tube is fixed to the skin next to the anus with 2 sutures.

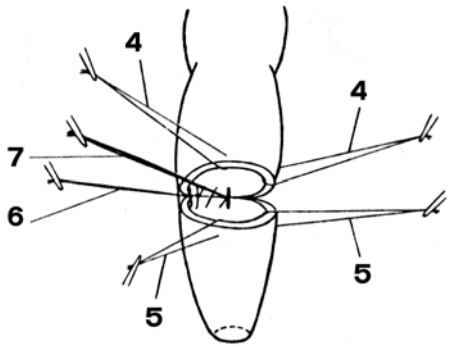

Fig. 3 The first quarter of the anastomosis (= right half of the posterior wall) is finished. (4) Holding suture at the edge of the divided descending colon, (5) holding suture at the edge of the rectal stump, (6) suture at right-hand corner, (7) suture in the middle of the posterior wall

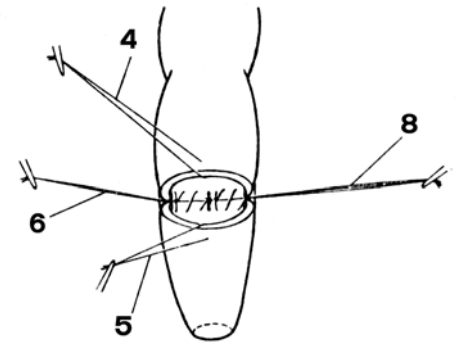

Fig. 4 Suturing of the posterior wall completed. (8) Suture at left-hand corner

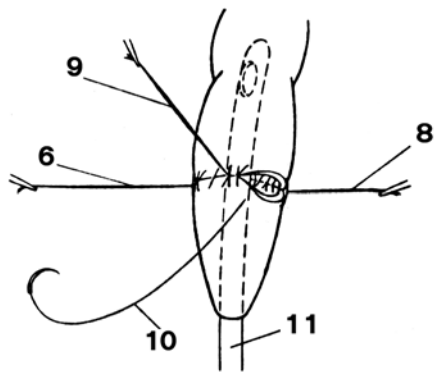

Fig. 5 Suturing of the right half of the anterior wall completed, (9) interrupted suture in the midline, (10) the last quarter of the anastomosis is sutured, (11) plastic bowel tube introduced

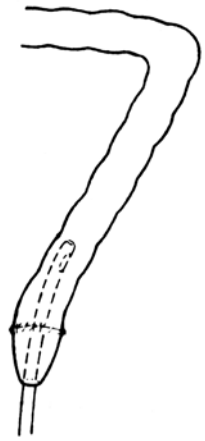

Fig. 6 The anastomosis is finished. The bowel tube lies above the anastomosis in the descending colon

9. Peritonealizing the Wound, Suturing the Abdominal Wall

The peritoneum at the rear is joined to the serosa of the descending mesocolon, using fine interrupted sutures. The abdominal cavity is closed with interruptd sutures.

Professor Fritz Rehbein evaluated the result of surgical treatment of Hirschsprung's disease into 4 categories[4]

Category*1*. *(Very good*): daily stool evacuation without any help

Category*2*. *(Good*): occasional laxative or enema

Category*3*. *(Satisfactory*): giving laxative or enema regularly

Table 1. Number of patients and sex during year 1987- 2000

year	87	88	89	90	91	92	93	94	95	96	97	98	99	00
boy	-	3	2	3	2	4	6	3	6	7	3	4	3	4
girl	1	2	-	1	2	-	2	2	2	1	3	5	2	-

Category*4. (Unsatisfactory*: no adequate bowel evacuation despite giving laxative or enema.

Follow-ups were scheduled for every two months in the first year and every four months in the second year and then every year after Rehbein's procedure.

Results

There were 50 boys and 23 girls (Table 1). The age of the patients at Rehbein's procedure varied from 7 months to 10 years (Table 2).

The length of the aganglionic segment included the rectosigmoid in 65 patients, the rectosigmoid and descending colon in 6 patients and the entire colon in 2 patients.

The result of follow-ups in 73 operated children with Rehbein's procedure, revealed very good results in 55 (= 75.3%), good results in 15 (=20.6%), satisfactory results in 2 (= 2.7%), and unsatisfactory results in 1 (= 1.4%). There was no death in the present series.

Postoperative complications was narrowing of the anastomosis in 20 patients, most likely to occur in cases where colostomy had to be performed in the newborn period. These conditions were treated by bouginage with hegar dilators in the hospital and continued as necessary by the parents at home. Postoperative enterocolitis occurred in 5 patients. Two patients with enterocolitis were treated successfully as out-patients, whereas 3 patients necessitated hospitalization and colonic decompression and antibiotics. Recurrent episodes of enterocolitis occurred in 2 patients.

Table 2. Age of patients at operations and number of colostomy and ileostomy

Age (year)	Number of Patient (n = 73)	Colostomy (n = 24)
0-1	11	-
>1-2	34	19 (ileostomy 2)
>2-3	7	2
>3-4	7	1
>4-5	5	1
>5-6	1	-
>6-7	-	-
>7-8	6	1
>8-9	1	-
>9-10	1	-
Median age	1-2	1-2

Dicussion

The results of Rehbein's procedure in Ratchaburi Hospital revealed very good in 55 (= 75.3%) and good in 15 (= 20.6%). When the patients with very good and good results are combined, the resulting figure is 95.9%. Professor Rehbein regard the main problem after surgical treatment of Hirschsprung's disease to be the gradual recurrence of constipation. The reappearance of disorders of stool evacuation cannot be attributed to inadequate resection of the narrow segment, leaving in aganglionic rectal tissue, but is due exclusively to the return of tenacious sphincter achalasia. Rehbein was convinced that the central problem in the treatment of Hirschsprung;s disease after the removal of the narrow segment was the permanent elimination of the sphincter achalasia[4]. A special collagenous fibrous layer called adventitia rectalis was found to limit the free expansion of the rectal wall. This tissue plays an important role in bowel movement by defining a threshold of intrarectal pressure in the bowel reflex of defecation and by transmitting the expulsive force from the abdomen above down to the anal opening. This discovery may explain why in Hirschsprung's disease, the preservation of the rectal wall in Rehbein's procedure will preserve better bowel function[11]. In conclusion, Rehbein's procedure is technically simple and yields good results. It should be another treatment of choice for Hirschsprung's disease.

Acknowledgements

The author wishes to thank Professor Polpatt Talalak, and Professor Chana Sathornkich, Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine, Siriraj Hospital, Mahidol University for their very useful advice.

References

1. Skinner MA. Hirschsprung's disease. Curr Prob Surg 1996; 33: 391-461.

2. Ryan ET, Ecker JL, Christakis NA. Hirschsprung's disease: associated anomalies and demographics. J Pediatr Surg 1992; 27: 76-81.
3. Lambrecht W, Kluth D, Gauderer MWL. In: Memoriam. J Pediatr Surg 1992; 27: 545-7.
4. Rehbein F, Booss D. Surgical Treatment of Hirschsprung's Disease. In: Holschneider AM, ed. Hirschsprung's Disease. New York: Thieme-Stration Inc, 1982: 189-99.
5. Sieber WK. Hirschsprung's Disease. In: Welch KJ, Randolph IG, Ravitch MM, O'Neil JA, Rowe MI, eds. Pediatric Surgery. 4th ed. Chicago: Year Medical Publishers, 1986: 995-1020.
6. Sangkhathat S. Congenital megacolon: New insight into molecular level. Thai J Surg 2000; 21(4): 113-20.
7. Swenson O. Hirschsprung's Disease. In: Raffensperger JG, ed. Swenson's Pediatric Surgery. 4th ed. New York: Appleton Century Croffs, 1980: 507-31.
8. Lister J, Rickham PP. Hirschsprung's Disease. In: Rickham PP, Lister J, Irving IM, eds. Neonatal Surgery. 2nd ed. London: Butterworth, 1978: 441-55.
9. Martin LW. Hirschsprung's Disease. In: Holder TM, Ashcraft KW, eds. Pediatric Surgery. Philadelphia: WB Saunder, 1980: 389-400.
10. Martin LW, Black RE. Colon and Rectum. In: Welch KJ, ed. Complications of Pediatric Surgery. Philadelphia: WB Saunder, 1982: 299-310.
11. Fu M, Zhang J: The discovery and a study of the adventitia rectalis, a fibrous layer of the rectal wall. J Pediatr Surg 1997; 32: 7-11.

การผ่าตัดวิธี Rehbein ในผู้ป่วยลำไส้ใหญ่โป่งพองแต่กำเนิด 73 ราย

สมชาย เทพเจริญนิรันดร์

หลักการรักษาโรคลำไส้ใหญ่โป่งพองแต่กำเนิดด้วยวิธี Rehbein คือการตัดลำไส้ส่วนที่เล็กซึ่งไม่มีเซลล์ประสาทรวมทั้งส่วนที่โป่งพองออกด้วยการเลาะเรคตัมส่วนบนลึกลงไปในอุ้งเชิงกรานให้ต่ำกว่าเยื่อบุช่องท้องประมาณ 2 เซนติเมตร และกำจัดการหดเกร็งของหูรูดส่วนในด้วยการขยายทวารหนักอย่างแรง ระหว่างเดือนสิงหาคม พ.ศ. 2530 ถึงเดือนเมษายน พ.ศ. 2543 มีผู้ป่วยโรคลำไส้ใหญ่โป่งพองแต่กำเนิด 73 ราย (เด็กชาย 50 ราย เด็กหญิง 23 ราย) ได้รับการผ่าตัดด้วยวิธี Rehbein ที่หน่วย ศัลยกรรมเด็ก โรงพยาบาลราชบุรี ผู้ป่วยเด็กมีอายุระหว่าง 7 เดือนถึง 10 ปี ผลการรักษา มีผู้ป่วย 55 รายสามารถถ่ายอุจจาระได้เองทุกวัน ผู้ป่วย 15 รายใช้ยาระบายหรือสวนทวารเป็นครั้งคราว ผู้ป่วย 2 รายต้องใช้ยาระบายหรือสวนทวารเป็นประจำ และผู้ป่วย 1 รายไม่สามารถถ่ายอุจจาระออกได้หมดทั้ง ๆ ที่ใช้ยาระบาย หรือ สวนทวาร โดยสรุปการผ่าตัดด้วยวิธี Rehbein ใช้เทคนิกง่ายผลการรักษาดีควรใช้เป็นอีกวิธีหนึ่งในการรักษาโรคลำไส้ใหญ่โป่งพองแต่กำเนิด