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Hirschsprung's Disease and Rehbein's Procedure – Our Results in the Last 30 Years

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A B S T R A C T

Hirschsprung's disease is congenital anomaly of the intestine and Harald Hirschsprung gave the first description of this disease[1]. The aim of this follow-up study was to evaluate the results of Rehbein's procedure in the treatment of Hirschsprung's disease in the last 30 years in Children's Hospital Zagreb. Hirschsprung's disease is congenital intestinal aganglionosis as the results of arrested fetal development of the myenteric nervous system. Hirschsprung's disease is affecting between 1:5000 to 1:8000 live births. A total of 124 children underwent Rehbein's lower anterior resection at Children's Hospital Zagreb. The principle of Rehbein' procedure is to remove aganglionic narrow segment and dilated sigmoid colon and anastomosis between normal intestine with rectal stump. The postoperative outcome was analysed for early and late complications like wound infections, abscesses, anastomotic insufficiency, postoperative enterocolitis, constipation, fecal incontinence, need for reoperation, ileus and mortality. On the basis of our results and data from literature we concluded that Rehbein's procedure is an excellent method for treatment Hirschsprung's disease.

Key words: *Hirschsprung's disease, Rehbein's procedure, children*

Introduction

Normal intestinal motility depends on a coordinated segmental contraction wave immediately preceded by smooth muscle relaxation as it propagates caudally. Patients with Hirschsprung's disease lack functional myenteric nervous system in the affected distal intestine and have ineffective distal peristalsis[2,3]. The clinical outcome are failure to pass meconium shortly after birth, failure to pass the first stool within 24 to 48 hours after birth, constipation, abdominal distension, palpable loops of bowel, vomiting,watery diarrhoea in the newborn, poor weight gain, slow growth and malabsorption. The aganglionic distal segment of the bowel is reason for dilatation of the proximal part of the colon or opening debility of the anal sphincter system[4,5].

The Rehbein's procedure was developed in 1953 by Fritz Rehbein, Germany[6]. This operative treatment offers few advantages: 1. No risk of damages autonomic nerve plexus of the small pelvis which supplies bladder, urethra, genital organs and the rectum. 2. Avoidance of incontinence. 3. Technical easy operation 4. Extraperitoneal location of the anastomosis[7].

The purpose of the present study was to present our results of treatment of Hirschsprung's disease with Rehbein's procedure.

Materials and Methods

The diagnosis of the Hirschsprung's disease was established on clinical signs, roentgen examination, anorectal manometry and rectal biopsy. In normal individuals, transient rectal distension causes relaxation of the internal anal sphincter. Anorectal manometry detects the absence of the relaxation reflex of the internal sphincter after the distension of the rectal lumen. In Hirschsprung's disease contraction occurs[8,9]. RTG examination with gastrografin enema shows us a spasmotic distal intestinal segment with dilation of the proximal bowel. This spastic transitional segment may be best seen on the lateral view. Findings in neonates are difficult to interpret because it often fails this transition zone, which takes time to develop. A suggestive finding is the failure to evacuate barium from the colon within 24

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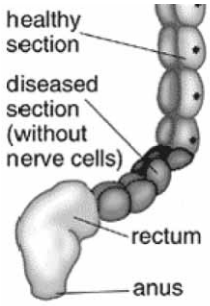

Fig. 1. Hirschsprung's disease.

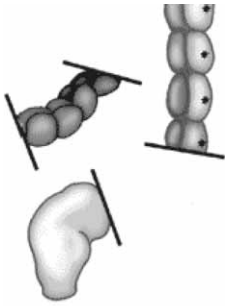

Fig. 2. – Removal of aganglionic intestine.

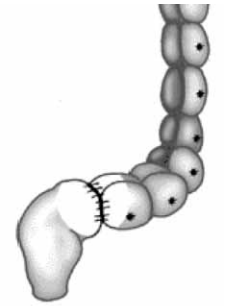

Fig. 3. End-to-end anastomos (normal colon+rectal stump).

hours of the performance of the study. The definitive method for obtaining tissue for pathologic examination is by a full-thickness rectal biopsy. Also simple suction rectal biopsy has been used to obtain tissue for histologic examination. The histologic and histochemical examinations suggest the absence of intramural ganglionic cells (submucosa Meissner's plexus and myenteric Auerbach's plexus) and the presence of excess nonmyelinated nerves in the distal intestine in an adequate rectal biopsy establishes the diagnosis[10,11]. The diagnosis of Hirschsprung's disease was confirmed during or after the operation. Rehbein's procedure removes aganglionic intestine (spasmotic part of intestine) with sigmoid colon and the dilated part of the intestine end-to-end anastomosis with rectal stump can be performed (Figures 1–3). The size of the rectal stump measures 3–4 cm in infants and small children and 5–7 cm in older children. A plastic tube was inserted through the anastomosis[11,12]. In patients with large colonic enlargement of the proximal part of intestine a right transverse colostomy was the procedure of choice for decompression. When we make definitive procedure we close the colostomy [13,14].

From 1974 to 2004 a total of 124 children underwent Rehbein's procedure and were followed up. The follow up period was 1 month to 12 years after operative treatment.

Results and Discussion

In the last 30 years 124 patients underwent Rehbein's procedure for treatment of Hirschsprung's disease. 97 (78.2%) patients were male and 27 (21.8%) patients were female. The sex ratio male to female was 3.6 to 1[15]. The follow up period was 1 month to 12 years after operative treatment and the average period was 4.5 years. The mean age of the patients at the time of operation was 4 years and 7 months, the range was 1 year to 17.8 year. 118 (95.1%) of patients were younger of 5 years at the time of operation.

Incidence of early complications was 8.1% (10 patients) wound infections, 10.5% (13 patients) were anastomotic stenosis, and 2.4% (4 patients) suffered anastomotic insufficiency. Only one patient (0.8%) died from toxic enterocolitic (Table 1).

In 44.4% (55 patients) preoperative colostomy was performed. One year after colostomy Rehbein's procedure was done. During the Rehbein's procedure closure of the colostomy was done in 72.7% (40 patients). 15 patients had one more operative treatment for closure colostomy. The most common complications of colostomy are prolapse or strictures. We have had no colostomy complications.

71.7% (89 patients) had stool every day, 13.7% (17 patients) had stool every second or third day and 14.5% (18 patients) showed rare bowel movement. The late postoperative complications were constipation in 12.9% (16 patients), intermittent diarrhea in 15.4% (19 patients) and fecal incontinence in 5.7% (7 patients). In total of 124 Rehbein's procedures 46% (57 patients) had recurrent achalasia of internal sphincter muscle and these patients required sphincter and anastomosis dilatation. In 95% of patients dilatations were successful and only 5% of these patients needed postoperative sphincterotomy. Stenosis of the anastomosis had 6.5% (8 patients) and these patients needs reoperation and reresection of these anastomosis. Only 1.6% (2 patients) had adhaesive ileus.

In total, excellent results means bowel evacuation every day, without any complications. Excellent results oc-

TABLE 1
COMPLICATIONS AFTER REHBEIN'S PROCEDURE

	Patients	Percentage
Wound infection	10	8.1%
Anastomotic stenosis	13	10.5%
Anastomotic insufficiency	4	2.4%
Toxic enterocolitis	1	0.8%

TABLE 2
LATE RESULTS AFTER REHBEIN'S PROCEDURE

	Patients	Percentage
Excelent	75	60.5%
Good	38	30.6%
Poor	11	8.9%

curred in 60.5% (75 patients). Good results had 30.6% (38 patients) and good results mean occasional laxative or enema for daily stool. Poor results had 8.9% (11 patients) and it means fecal incontinence or laxatives and enema were given regularly for stool (Table 2).

Conclusion

The world surgical literature describes different surgical procedures for the definitive surgical treatment of aganglionosis or Hirschsprung's disease. Every method involves bringing normal bowel as low in the rectum as is technically possible by resecting or bypassing the aganglionic bowel[16–18]. Rehbein's procedure removes aganglionic intestine (spasmotic part of intestine) and the dilated part of the intestine end-to-end anastomosis with rectal stump can be performed. Rehbein's procedure does not disturb the sphincter mechanism. Sigmoid colon was always resected because in this part of colon the water was absorbed. Without sigmoid colon stool was fluent and passed easily by through rectal stump. The size of the rectal stump measures 3–4 cm in infants and small children and 5–7 cm in older children. The technique is simple and can be used for treatment even in early infancy. We avoid preoperative colostomy when possible and make primary definitive operation. In our hospital we perform only Rehbein's procedure for treatment of Hirschsprung's disease. Due to the excellent and good results achieved in 91.1 % of the patients, we regard Rehbein's method excellent for the treatment of Hirschsprung's disease[19,20].

In the last year we performed a laparoscopic Duhamel pull-through method for Hirschsprung' disease. If it is not possible to perform a laparoscopic method we perform Rehbein's procedure[21–25].

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HIRSCHSPRUNGOVA BOLEST I REHBEINOVA METODA LIJEČENJA – NAŠI REZULTATI U ZADNJIH 30 GODINA

SAŽETAK

Hirschsprungova bolest je urođena anomalija crijeva, a Harald Hirschsprung je prvi opisao tu bolest[1]. Svrha ovog rada je evaluacija rezultata operacije Hirschsprungove bolesti po metodi Rehbein u zadnjih 30 godina u Klinici za dječje bolesti Zagreb. Hirschsprungova bolest je urođena bolest crijeva koja nastaje radi zastoja u fetalnom razvoju myenteričkog nervnog sustava. Sama bolest zahvaća jedno od 5,000 do 8,000 živorođene djece. Ukupno je operirano 124 djece po metodi Rehbein u našoj bolnici. Osnovna zamisao ovog operativnog zahvata je odstranjenje uskog dijela crijeva bez ganglijskih stanica zajedno sa sigmom te spajanje zdravog crijeva sa ostatkom rektuma. Postoperativni rezultati su evaluirani kao rane i kasne komplikacije i to kao upala rane, gnojni procesi u trbuhu, slabost ili suženje učinjene anastomoze, postoperativni učestali proljev ili zatvor, nemogućnost kontrole stolice, potreba za ponovnom operacijom, ileus te smrtni ishod liječenja. Na osnovu naših rezultata i podataka iz literature možemo zaključiti da je metoda po Rehbeinu odlična za liječenje Hirschsprungove bolesti.