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ORIGINAL ARTICLE

A 15-Year Experience with the One-Stage Surgery for Treatment of Hirschsprung's Disease in Newborns, Infants, and Young Children

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Abstract The treatment of Hirschsprung's disease has changed over the past several years. Significant modifications occurred after the implementation of surgery without laparotomy, using transanal access. The type of this surgery depends on the condition and the age of a child. The aim of the study was to summarize our 15-year experience with one-stage surgery for the treatment of Hirschsprung's disease in the wider context of current clinical practice and to identify transanal endorectal pull-through-related factors influencing the surgery and hospitalization. The retrospective analysis of newborns, infants, and young children was performed between 2000 and 2014. Four girls and 29 boys were operated on. The parameters describing the surgery and the hospitalization were analyzed. The number of patients treated using transanal endorectal pull-through technique was 30, and Duhamel-Martin, 3. There were significant correlations ($p<0.05$) between necessity of blood transfusion, length of resected intestine, operative time, the number of intraoperative histopathological assessments, and length of hospitalization. The time of one-step surgery is extended because of waiting for repeated intraoperative histopathological assessment of the level of resection. More extended bowel resection is connected with longer length of hospitalization and, more often, necessity of blood transfusion.

Keywords Child · Congenital intestinal aganglionosis · Hirschsprung's disease · Surgical procedures

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Introduction

Hirschsprung's disease is a congenital disorder caused by abnormal innervation of the intestine, as a result of the failed migration of neuroblasts [1]. This results in disturbances of peristaltic wave conduction and leads to intestinal obstruction.

Treatment of Hirschsprung disease is surgery. The pathological segment of the bowel is removed. Then the anastomosis is made between properly innervated intestine and the anus, above the external sphincter. The surgery can be performed in a single step, which is a preferred method of treatment, or in stages using temporary intestinal stoma [2]. The choice of surgical procedure depends on the clinical condition of the patient, the severity of symptoms, and the age of child [3]. Actually, surgical methods include Swenson, Soave, Duhamel, or Rehbein techniques used in different modifications [4, 5].

The Swenson Procedure

The patient is positioned on the operating table to provide simultaneous exposure of the perineum and abdomen. The pelvis is allowed to drop back over the lower end of the table and legs are strapped over sandbags. The Swenson technique includes aganglionic rectosigmoidectomy with end-to-end anastomosis of normal colon just above the anal sphincter. The rectum is released from perirectal tissues by precise dissection around the rectum down to the anal canal. The mobilized aganglionic rectum and anal canal are everted, and anastomosis between the pulled-through ganglionic colon and anal canal is performed outside the anus [6].

The Soave Procedure

The patient is placed into lithotomy position. This surgery requires abdomino-perineal access. Separation between the

seromuscular and muscular layer is performed. The colon is pulled down and is passed through the rectal canal. The aganglionic segment of the intestine is removed by pulling through the muscular cuff outside. The normal ganglionic colon is brought, and coloanal anastomosis is performed [7].

The Duhamel Procedure

The Duhamel-Martin technique is carried out in two phases: abdominal and perineal. In the abdominal phase, the aganglionic and transition zones of the gut are identified and mobilized to pelvic peritoneal reflection. The normally innervated gut is placed in created retrorectal space and pulled through the posterior rectal wall and anus. Then an end-to-side and side-to-side anastomoses are made between the aganglionic rectum and normal intestine using circular and longitudinal stapling devices. Finally, the abnormal gut is resected.

Initially, biopsies are taken before mobilizing the sigmoid colon and rectum. The final decision on the extent of resection upwards is made according to the results of histological examination [2, 8].

The Rehbein Procedure

The Rehbein method includes removal of the aganglionic colon up to the upper rectum followed by a vigorous dilatation of the rectum and anal canal. Actually, this procedure is performed rarely [9].

Transanal Endorectal Pull Through

New possibilities are actually created by a technique of removing the aganglionic intestine through the anus-transanal endorectal pull through (TEPT). This method was described by De la Torre and Ortega in 1998, and since this moment, this type of surgery rapidly spread abroad, in many centers of pediatric surgery [10]. It involves complete dissection and mobilization of the entire aganglionic colon and anastomosis of the normal colon through the anus. Transanal access bases on traditional surgical techniques performed previously in abdominal approach. The Soave and Swenson modifications are commonly used [11, 12]. This type of surgery is used for the treatment of small children [13].

Transanal endorectal pull through using the Soave technique is carried out with lithotomy position. It involves removal of rectal mucosa submucosa and pulling through the ganglionic intestine through the muscular cuff of the aganglionic intestine. After eversion of the intestine, the dissection of mesenteric vessels is performed. The next step of surgery is uptaking biopsy specimen from the intestine, on the level which seems to be normally innervated, based on an earlier radiological examination [10]. After confirmation of the presence of normal ganglion cells in the intestine, the affected bowel is resected and anastomosis is done between the normally innervated bowel and the anal canal above the dentate line.

TEPT using the Swenson technique is a transanal full-thickness dissection of the rectum. After entering the abdominal cavity, other steps are the same as in the Swenson procedure described above. Surgery is performed in a prone buttock-elevated position of the patient.

Over the past 15 years, the popularity of minimal-access surgical techniques has led to a number of changes to the standard single-staged operations, including the use of laparoscopic access [14].

The aim of the study was to summarize our 15-year experience with one-stage surgery for treatment of Hirschsprung's disease in the wider context of current clinical practice and to identify transanal endorectal pull-through-related factors influencing the course of the surgery and hospitalization in the group of neonates, infants, and young children.

Materials and Methods

The material for the study was collected on the basis of documentation of patients treated in the Department of Pediatric Surgery, Traumatology and Urology in Poznan over the years from 2000 to 2014. There were treated 4 girls and 29 boys using one-stage surgery methods: TEPT (30 patients) and Duhamel-Martin (3 girls).

Transanal endorectal pull-through procedure using the Soave technique was performed in 29 patients and the Swenson modification in 1 child. The Swenson method in TEPT technique has been used in our institution since 2013.

Additionally, the abdominal approach was required in two patients, who were treated with Soave's transanal endorectal pull-through surgery. In these children, 38 and 61 cm of the intestine were resected. A short laparotomy incision made it possible to mobilize the descending colon. Detailed description of the techniques was presented in the "Introduction."

In all these cases, earlier preoperative evaluation was carried out on the basis of contrast enema. The result of this examination decided about the planned length of intestinal resection.

For each case, preparing the patient for surgery included full colon cleansing with a washout enema until clear, performed under general anesthesia, just before the operation.

Prophylactic antibiotic therapy conforming to current internal guidelines issued by the hospital was initiated on the day of the surgery and continued during hospitalization.

The analysis included the following: age of patient, length of resected intestine, operative time, necessity of blood transfusions during and after surgery, number of intraoperative

histopathological assessments, and length of the hospitalization after surgery.

The results were collected so that evaluations of distributions of assessed variables and analyses of their parameters could be conducted and patients could be grouped with respect to analyzed characteristics. Calculations were performed using STATISTICA 10 (StatSoft Inc., Tulsa, USA). The significance level was $\alpha=0.05$. The result was considered statistically significant when $p<\alpha$. To examine the relationship between variables measured on an interval scale, due to the lack of compliance with the normal distribution, the Spearman's rank correlation coefficient was calculated. For comparison of variables, which were measured using an interval scale and which had the normal distribution, the Student's t test was used for unrelated groups (in the case of equality of variance) and the Cochran-Cox test was used (no homogeneity of variance). For the variables, which were not normally distributed, the Mann-Whitney test was used.

The design and methods used in the study were approved by the Bioethical Commission of Poznan University of Medical Sciences (Decision No. 58/12).

Results

Analysis of the results showed that the average age of patients operated on using the TEPT technique was 12 months. This method was used in children aged from 3 weeks to 7 years: 12 neonates, 12 infants, and 6 young children. The Duhamel-Martin technique was used in three girls at the age of 8 months, 4 and 5 years.

The average length of resected intestine in the TEPT method was 22 cm; the shortest and the longest lengths of the resected bowel were 12 and 61 cm. In patients treated using the Duhamel-Martin technique, segments 15, 18, and 20 cm of the intestine were resected.

The mean time of surgery was 3 h and 20 min for the TEPT method. The shortest and the longest times of this operation were 2 and 5 h. Girls treated using the Duhamel-Martin surgeries were operated on for 2 h.

Six children needed blood transfusion during or just after surgery. Intestinal resection in these patients ranged from 15 to 60 cm. Loss of blood caused by a long-lasting surgery or difficult dissection and amount of blood, which was lost in the resected bowel, affected a necessity of transfusion.

Twenty-eight patients had a single intraoperative histopathological assessment. Other five patients had two or three examinations during surgery.

The average time from surgery to discharge the child from the hospital was 11 days for the TEPT method. The shortest and the longest times of hospitalization were 4 and 23 days. Children operated on using the Duhamel-Martin technique were hospitalized after surgery for a period of 8, 16, and 17 days.

Correlations between the above-mentioned elements were found in the study.

In the group of children operated on Soave's transanal endorectal pull-through surgery, a correlation between the length of the resected bowel and length of the hospitalization was found: children, who had longer resection of the intestine, had longer hospitalization after surgery (Fig. 1).

The analysis of the results also found the correlation between necessity of blood transfusion, length of the resected intestine, operative time, and length of the hospitalization after surgery.

Children, who had blood transfusion, in comparison to patients without this, had longer resection of the intestine, longer operative time, and length of the hospitalization (Figs. 2, 3, and 4).

Children, who had two or three intraoperative histopathological assessments, had longer operative time. The time of surgery of patients who were operated on for 4 h and longer was connected with the necessity of repeated intraoperative histopathological evaluation. The first intraoperative histopathological examination, in five cases, showed abnormal aganglionic structure of the bowel. In these cases, longer resection and again histopathological examination were performed (Fig. 5).

Discussion

Transanal endorectal pull-through method performed with transanal access is characterized by low invasiveness of surgery and good results of treatment [15–17].

In our institution, the TEPT technique was introduced in 2004, and it was the first center in Poland operating on children by using this method [18]. Currently, this method is used in most centers treating Hirschsprung's disease in Poland.

Czauderna et al. highlight the benefits of this technique in his research. They describe TEPT as an effective and minimally invasive technique. During surgery, there was no need to open the abdominal cavity. This allowed for earlier implementation of oral feeding, better cosmetic results, and shorter hospital stay [4].

The TEPT method is the preferred method of treatment, especially of smaller children with uncomplicated form of Hirschsprung's disease. A necessary condition to qualify a patient for a single-stage surgery is lack of symptoms of active enterocolitis.

The transanal endorectal pull-through technique adapts the Soave, Swenson, or Rehbein procedure during surgery. In most of cases in our department, the Soave procedure was used. This modification is often used, because of its good

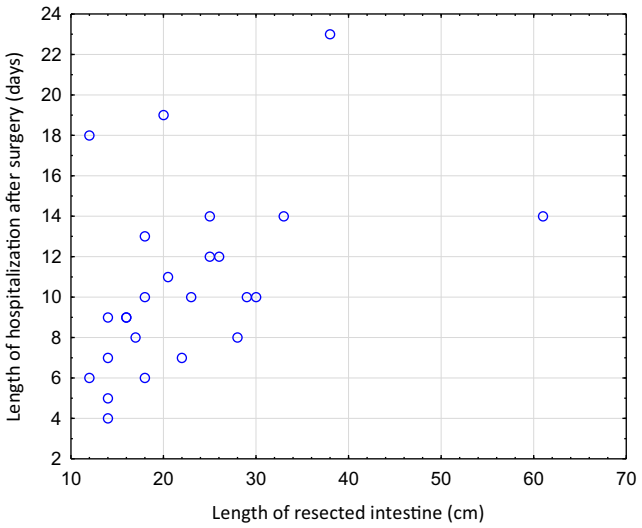

Fig. 1 Scatterplot illustrating the correlation between the length of resected intestine and the length of hospitalization in patients operated on using Soave's transanal endorectal pull-through surgery

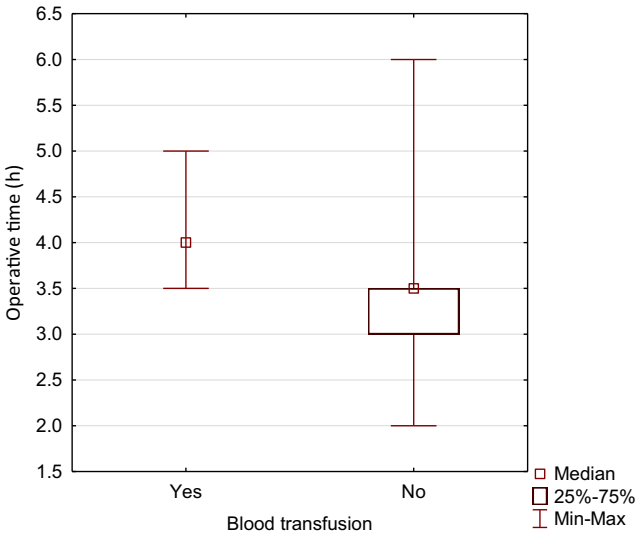

Fig. 3 Boxplot presenting the lower median operative time in patients who did not require blood transfusion during or after Soave's transanal endorectal pull-through surgery

results reported after treatment, both with and without use of colostomy [19].

The Swenson method in TEPT modification has been used in our institution since 2013. Advantages of this method were described by Levitt et al. who claim that Swenson-like dissection technique is safe, reproducible, and avoids many of the long-term complications associated with the other techniques [20].

Older children with diagnosed short-segment form of Hirschsprung's disease have less severity of symptoms. Reduced alertness of parents because of scant symptoms, delayed pediatric care, and delayed diagnosis may affect on the final late treatment of the disease.

The analysis of the results of the study did not prove the correlation between the age of operated patients and the length of their aganglionic intestine. The shorter length of the resected intestine was not related to the older age of the patient. It is probably caused by an earlier detection of even scant signs of the short-segment type of Hirschsprung's disease and earlier, more precise diagnostics. In this situation, both patients with short-segment form and long-segment form of Hirschsprung's disease are diagnosed at young age. The increasing knowledge of Hirschsprung's disease among the medical community also has influence on the earlier detection of the disease.

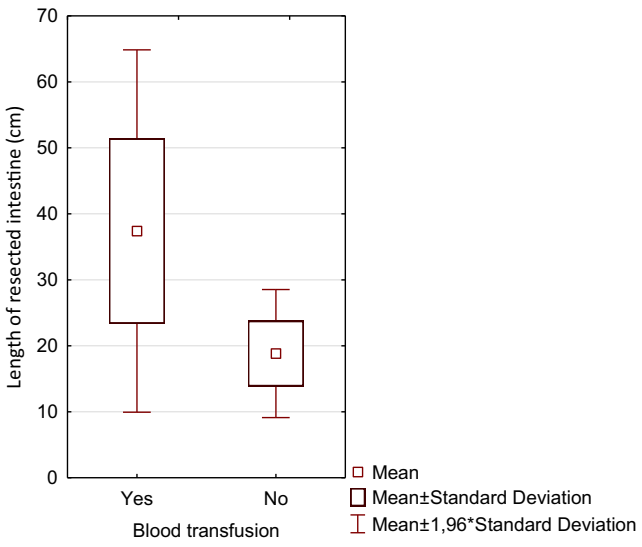

Fig. 2 Boxplot comparing the length of the resected intestine in patients operated on using Soave's transanal endorectal pull-through surgery, in those who necessitated blood transfusion and in those who did not require it

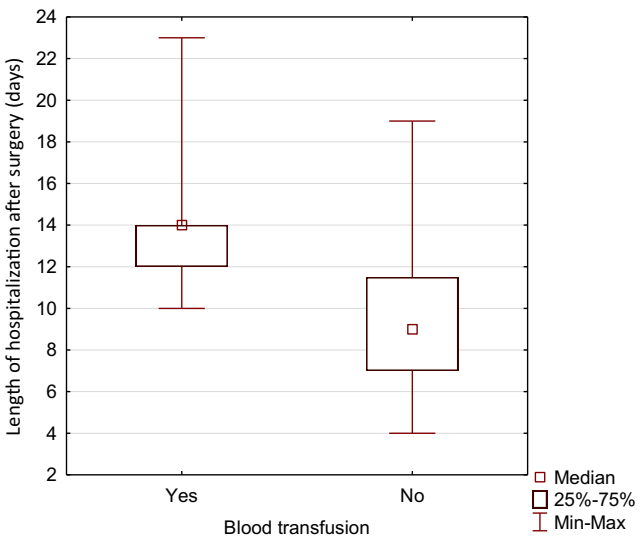

Fig. 4 The length of hospitalization after surgery depending on the necessity of blood transfusion during or after Soave's transanal endorectal pull-through surgery

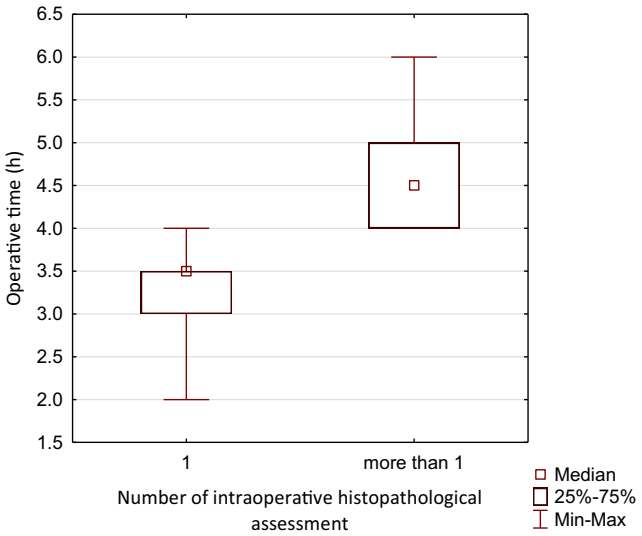

Fig. 5 The boxplot illustrates the influence that additional intraoperative histopathological analyses have on the operative time in patients operated on using Soave's transanal endorectal pull-through surgery

Even though described children might be operated on using the Duhamel-Martin technique, they were treated using the TEPT surgery. Factors promoting the choice of this technique were present in these children. The segments of the aganglionic intestine were short, which allows for safe dissection and mobilization. It is also noteworthy that parents often express greater satisfaction with the one-stage technique through the anus because of better cosmetic effect. In this aspect, our experience is consistent with the findings presented by Georgeson et al. [21].

The principle of the Duhamel procedure is partially bypassing the rectum and performance of anastomosis end to side. Abdominal dissection also may be completed during laparoscopy. Surgery is performed using linear and circular stapler. The size of disposable staplers limits the possibilities of their use. In small children, the only option for this type of operation is using laparoscopic linear staplers, due to their small size, but we did not dispose of such a circular stapler [22].

Children with the long-segment form of the Hirschsprung's disease were hospitalized longer after operation. It was caused by a more difficult surgery and, therefore, necessity of longer observation of them.

The presented data suggest that the length of the resected fragment of the intestine influenced the blood loss. Patients with long-segment Hirschsprung's disease more often have the necessity of the blood transfusion during or after surgery. It was caused by the amount of blood, which was lost in the resected intestine. The necessity of blood transfusion also was connected with longer duration of the surgery. It is caused by the loss of blood during long or difficult dissections.

If the time of surgery was longer, it was associated with a loss of blood and necessity of transfusion. Transfusion was performed during or after surgery depending on the values of hematological parameters. After transfusion, it was safer to observe the child for a longer period of time and perform additional controls of laboratory parameters in the inpatient setting. Although such observation could elongate the hospitalization, in our experience, the need for a longer stay in cases of longer resections is related to the procedure itself, i.e., to the healing of the anal region.

In every case, the contrast enema was performed before surgery. This examination may show a view of the type of Hirschsprung's disease. Unfortunately, the form of disease showed in the contrast enema does not always correlate with the result of intraoperative histopathological evaluation of the resected intestine. In the transanal approach, the level of intraoperative biopsy is estimated basing on preoperative radiological examination. When the result of biopsy is aganglionic or transitional zone, the next resection and repeated histology is needed. The time of waiting on the result of this examination additionally elongates the time of the entire transanal endorectal pull-through surgery. During surgery with abdominal access, biopsy is made at the beginning of the operation, which causes that waiting for histopathological evaluation does not elongate the time of surgery significantly [23].

The Duhamel-Martin technique is used in older patients. The appropriate size of the patient's pelvic compartment allows using the staplers. It is also a much more invasive surgery because it requires opening the abdominal cavity [22]. In the Department of Pediatric Surgery, Traumatology and Urology in Poznan, the Duhamel-Martin technique is usually used as a second stage in treatment of Hirschsprung's disease.

Elhalaby et al. found in older children difficulties in preparation between mucosal and muscular layers during surgery using the Soave technique [24]. These limitations seem to be insignificant for the transanal operation using the Swenson technique [20]. Our initial experience confirmed this opinion.

Less invasive surgical techniques enable surgeon to achieve a high rank of success. It is worth paying attention to their development. They are characterized by good treatment effects while being less invasive for the child. Therefore, in case of uncomplicated form of Hirschsprung's disease in newborns, infants, and young children, the technique of choice is transanal endorectal pull through. Additionally, the above-mentioned factors may have an influence on the choice of surgical technique.

In conclusion, the most commonly performed surgery in the one-stage treatment of Hirschsprung's disease is the transanal endorectal pull through.

The transanal endorectal pull-through technique is characterized by a short period of hospitalization after surgery. The time of one-step surgery using this method is extended because of waiting for the result of intraoperative histopathological test of the resected intestine.

More extended resection of the intestine is related to longer duration of hospitalization and greater frequency of blood transfusion necessity.

Contrast enema does not allow for determination of the optimal level of resection.

Conflict of Interest The authors have no conflict of interest.

Ethical Standards All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. All persons gave their informed consent prior to their inclusion in the study.

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