Research Article

Surgical Correction of Hirschsprung’s Disease in Children Using the Soave-Boley Technique with Manual Colorectal Anastomosis

Oleh Kurtash

Abstract

Introduction. Current trends in surgical treatment of Hirschsprung’s disease are aimed at minimally invasive interventions. However, the experience of using Soave-Boley procedure in surgical treatment of Hirschsprung’s disease in children of different ages is valuable in the arsenal of differentiated approach to the treatment of this pathology.

The objective of the research was to evaluate the results of surgical correction of Hirschsprung’s disease in children using the Soave-Boley technique with manual colorectal anastomosis.

Materials and Methods. The analysis of surgical treatment of 1, 187 children with different forms of Hirschsprung’s disease aged from birth to 18 years over the period 1980-2020 was conducted in the National Children’s Specialized Hospital “Okhmadtyt”. Surgical correction of Hirschsprung’s disease using the Soave-Boley technique was performed in 597 children. Before surgery, 156 patients underwent the first stage of treatment that consisted in the creation of a protective colostomy; in 441 cases, this intervention was performed without an intestinal stoma; the benefits of the Soave-Boley technique were evaluated.

Results. All the patients survived. In 15 (2.51%) out of 597 children, in the early postoperative period, there were observed: retrocolic hematoma (n = 2), retrocolic abscess (n = 7), anastomotic leak (n = 2), adhesive intestinal obstruction (n = 3), intussusception (n = 1). Fourteen (2.36%) patients developed surgical complications in the long-term period: residual aganglionosis (n = 9), anastomotic stenosis (n = 4) and coloptosis. Repeated Soave-Boley operation with manual colorectal anastomosis was successfully performed in 26 (4.362%) patients after primary correction of Hirschsprung’s disease using different methods. Periodic episodes of fecal smearing in the remote period in 45 (7.53%) children were eliminated by conservative treatment. The success of this technique is confirmed by a much lower number of early (2.51%) and late (2.36%) postoperative surgical complications, as compared to those after using other methods of open surgical correction - 17.52% and 16.35%, respectively.

Conclusions. Surgical correction of Hirschsprung’s disease in children using the Soave-Boley technique with manual colorectal anastomosis allows better control over applying each suture, anatomical joining the edges of the bowel, and reducing trauma to the rectal mucosa, which ensures the high reliability of colorectal anastomosis; it is the most effective way of radical correcting Hirschsprung’s disease in children in an open way in one-stage or two-stage interventions, and this technique is the operation of choice for secondary surgical correction of Hirschsprung’s disease.

Keywords

Hirschsprung’s Disease; Children; Surgical Treatment; Results

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Hirschsprung’s disease (HD) is a complex malformation of the colon that may be treated only surgically. The essence of surgical correction is performing a resection of the aganglionic segment and a part of the altered intestine and imposing a colorectal anastomosis [2, 7].

Various methods of radical HD correction have been evolutionarily improved. The so-called open methods proposed by O. Swenson (1948), W. Duhamel (1956), F. Rehbein (1959), F. Soave (1963), H. Lynn (1956) and their numerous modifications appeared in chronological order [1, 6, 8, 14]. Each of these techniques has a specific goal in eliminating the causes and consequences of intestinal aganglionosis. As a result, the technique that is the least traumatic, the most technically acceptable, which eliminates all the pathogenetic factors and leads to the best functional results in the early and remote postoperative periods is acceptable for practical use. All these characteristics should certainly be accepted as a whole [9, 11, 16].

Current trends in surgical treatment of HD are aimed at minimally invasive interventions. K.E. Georgeson proposed one-stage laparoscopy-assisted surgical treatment of HD. De La Torre described the first results of his own method, namely transanal endorectal pull-through (TEPT) [13, 14].

However, in some cases, open techniques are not rejected by pediatric surgery [3, 15]. The Soave procedure modified by Boley is one of the most physiological and most technically acceptable technique among pediatric surgeons from different countries [5, 17]. Moreover, the main technical elements of this operation were included in the procedure of minimally invasive intervention in the correction of HD, namely TEPT. In addition, the Soave-Boley technique is a rational solution in conversion of minimally invasive interventions in patients with HD [4, 10, 12].

Therefore, the experience of using the Soave-Boley procedure in surgical treatment of HD in children of different ages is valuable in the arsenal of differentiated approach to the treatment of this pathology.

The objective of the research was to evaluate the results of surgical correction of HD in children using the Soave-Boley technique with manual colorectal anastomosis.

### Materials and Methods

The analysis of surgical treatment of 1,187 children with various forms of HD at the age from birth to 18 years over the period 1980-2020 was conducted (Table 1).

All the patients underwent both traditional and minimally invasive radical surgery. Fourteen children received the Swenson’s pull-through operation; 29 children underwent the Duhamel procedure; 7 children were operated on according to the Rehbein’s method; 59 children underwent the Soave procedure; 722 children underwent the Soave-Boley procedure. There was one more group which included 105 patients who underwent reconstructive plastic surgery in extensive forms of aganglionosis. Lynn’s sphincteromyotomy was performed in 42 cases of ultra-short HD.

In addition, since October 2011, we have operated 209 children with HD using a minimally invasive technique. Among them, 144 patients were operated on according to TEPT approach and 65 children were operated on according to laparoscopy-assisted TEPT approach.

To diagnose and evaluate treatment outcomes during postoperative monitoring, the results of clinical examinations and special methods of investigation were used. They included radiological (irrigography, irrigoscopy, contrast radiography of the digestive tract) and endoscopic methods (sigmoidoscopy, colonoscopy, anorectal manometry), functional (anorectal manometry) and morphological studying (histological study, determination of acetylcholinesterase activity).

Having analyzed a 40-year experience in surgical treatment of HD according to different methods retrospectively, we considered necessary to study three periods, namely 1980 to 1992, 1993 to 2010 and 2011 to 2020 each of which led us to approaches to improving the methods of surgical treatment of HD. It has become possible to modernize the methods of surgery, as well as to intensify the diagnostics for early detection of patients with HD (Table 2).

During the first period, we used so-called “open” surgeries, namely the Swenson, Duhamel, Soave procedures. During this period of our research, in the literature, there was a contentious debate over advantages and disadvantages of certain open methods. Therefore, we used different methods trying to reach a compromise in the differentiated approach to the selection of surgery.

In the second period, the main radical surgery we used was the Soave-Boley procedure modified by us with the implementation of manual or mechanical primary colorectal anastomosis (PCRA).

Since October 2011 (the third period), we have used minimally invasive techniques of HD surgical correction, namely TEPT and laparoscopy-assisted TEPT approach continuing to perform the Soave-Boley procedure in older children and those with a protective colostomy created during the first stage of surgery.

Reconstructive plastic surgery in extensive forms of aganglionosis and the Lynn procedure in uncomplicated ultra-short forms of HD, as methods of correcting a separate category of patients, were performed during all study periods and did not depend on the main principles of division into these groups.

### Ethical Statement

The research was carried out following the principles of the Declaration of Helsinki. The research protocol was approved by the Local Ethics Committee of Pediatric Surgery Clinic of the National Children’s Specialized Hospital "Okhmatdyt". Informed parental consent was obtained for the research.
Table 1. Distribution of patients according to age and type of aganglionosis.

<table>
<thead>
<tr>
<th>Type of aganglionosis</th>
<th>Patients’ age</th>
<th>Total</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0-6 months</td>
<td>6-12 months</td>
<td>1-3 years</td>
</tr>
<tr>
<td>Rectal</td>
<td>39</td>
<td>61</td>
<td>74</td>
</tr>
<tr>
<td>Rectosigmoid</td>
<td>98</td>
<td>94</td>
<td>187</td>
</tr>
<tr>
<td>Subtotal</td>
<td>102</td>
<td></td>
<td>7</td>
</tr>
<tr>
<td>Total colonic</td>
<td>41</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>280</td>
<td>203</td>
<td>268</td>
</tr>
</tbody>
</table>

Table 2. Distribution of patients according to variant of radical surgery during different periods of treatment modernization.

<table>
<thead>
<tr>
<th></th>
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</tr>
</thead>
<tbody>
<tr>
<td>Swenson procedure</td>
<td>14</td>
<td>-</td>
<td>-</td>
<td>14</td>
</tr>
<tr>
<td>Duhamel procedure</td>
<td>29</td>
<td>-</td>
<td>-</td>
<td>29</td>
</tr>
<tr>
<td>Rehbein procedure</td>
<td>7</td>
<td>-</td>
<td>-</td>
<td>7</td>
</tr>
<tr>
<td>Soave procedure</td>
<td>59</td>
<td>-</td>
<td>-</td>
<td>59</td>
</tr>
<tr>
<td>Lynn procedure</td>
<td>14</td>
<td>21</td>
<td>7</td>
<td>42</td>
</tr>
<tr>
<td>Manual Soave-Boley procedure</td>
<td>-</td>
<td>344</td>
<td>253</td>
<td>597</td>
</tr>
<tr>
<td>Soave-Boley procedure (by means of surgical stapler)</td>
<td>-</td>
<td>125</td>
<td>-</td>
<td>125</td>
</tr>
<tr>
<td>Reconstructive plastic surgeries in extensive forms of aganglionosis</td>
<td>1</td>
<td>90</td>
<td>14</td>
<td>105</td>
</tr>
<tr>
<td>TEPT</td>
<td>-</td>
<td>-</td>
<td>144</td>
<td>144</td>
</tr>
<tr>
<td>Laparoscopy-assisted TEPT</td>
<td>-</td>
<td>-</td>
<td>65</td>
<td>65</td>
</tr>
<tr>
<td>Total</td>
<td>124</td>
<td>580</td>
<td>483</td>
<td>1,187</td>
</tr>
</tbody>
</table>

Results and Discussion

The main technical elements of operations to restore the patency of the intestinal tract in surgical treatment of children with HD are the mobilization of the colon and rectum and the formation of colorectal anastomosis. Modernizing these elements simplifies surgery, makes surgery less traumatic, facilitates the postoperative period, accelerates the rehabilitation period, and improves the patients’ quality of life in the long-term period. Among the methods of open surgical management of HD, we prefer the Soave procedure modified by Boley as the most physiological and the most acceptable technique for the formation of manual colorectal anastomosis introduced in the Pediatric Surgery Clinic of the National Children’s Specialized Hospital "Okhmatdyt". Gradually, the formation of mechanical (stapler) colorectal anastomosis was added to this technique. Since special equipment (circular stapler) is required to perform stapler entero-enteroanastomosis, manual colonic anastomosis was the most common in the clinic.

Manual Soave-Boley surgery was performed as follows: before the procedure, patients were placed in the lithotomy position; the abdominal cavity was opened with a wide midline incision up to the suprapubic region; a thorough examination of the abdominal organs was performed; the entire colon was examined; its aganglionic portion and suprastenotic part were visualized; the state of the enlarged intestine and the proximal margin of colon resection were determined.

During the examination of the colon, in most patients, the aganglionic area differed distinctly in diameter as compared to the area of the colon located above this site. The aganglionic area was pink, with longitudinal folds and sclerotic thickened wall. The state of suprastenotic colonic dilation depended on the length of aganglionosis, the degree of narrowing, the patient’s age, the presence and severity of concomitant inflammatory process of the intestine.

If it was possible to achieve a good bowel movement before radical surgery, the area of suprastenotic dilation in such children was shorter, narrower, with more pronounced haustration and less hypertrophied intestinal walls in this area. Secondary changes in the supraganglionic zone were more pronounced in aganglionosis involving longer segments (above the level of the rectum), or after a long, ineffective conservative emptying of the intestine from fecal masses, or in case of protracted diagnosis of this pathology. The large intestinal segment, proximal to the narrowing site, was much wider, more haustric and sclerotic. This segment was pale pink; its mesentery was thinned, and stagnant purple blood vessels were significantly dilated.
We mobilized the colon from the marking site to the transi-
tional fold of the peritoneum, taking into account the archi-
tecture of the blood vessels and ureters.

Using the Soave technique, we separated a seromuscular
cuff 1-3 cm above the transitional fold of the peritoneum
(depending on the child’s age).

In children, who first underwent protective colostomy
creation, the seromuscular cuff was separated using the Soave
technique. Then, we separated the colon stump in the form
of a stoma from the anterior abdominal wall. The segment
of the colon needed for constructing and anastomosis was
mobilized.

The formation of the seromuscular cuff is the most difficult
stage of surgery. Technical difficulties during this stage oc-
curred due to pronounced sclerotic changes in the submucosal
layer of the aganglionic zone that usually resulted from pro-
longed mechanical release of the intestine from fecal masses
(the site of enema tip pressure), multiple dilations of the agan-
glionic area or prolonged influence of fecal masses before
mechanical cleaning.

Significant sclerotic changes in the rectum sometimes led
to damaging the mucous membrane during demucosation,
which required careful treatment with antiseptics and sutur-
ing the defect. Demucosation was performed at the level of
the dentate line of the rectum. Along the edges of the cuff,
there were placed 2 clamps, between which the muscular layer
of its posterior wall was dissected.

After that, the second surgical team of three people worked
simultaneously to perform perineal surgery.

In children older than three years, the retrocolic space
was drained post-sphincterically through a separate skin
incision using a PVC tube. The drainage was placed between
the inner wall of the retrocolic space and the posterolateral
surface of the rectum. Its introduction and placement were
controlled visually from the side of the abdominal wound.
The drainage was fixed with a separate ligature at the site
of skin incision on the perineum. The purpose of draining
the retrocolic space was to eliminate the exudate and possible
hemorrhage from the site of the demucosed cuff. Patients
older than three years are more likely to suffer from these
fluid formations. The separation of the seromuscular cuff in
children of this age group is the most traumatic. There is no
need to drain the retrocolic space in children under three years
of age.

After treating the lumen of the rectum with antiseptics,
we inserted a fenestrated clamp into the anus, to which a loop
which required careful treatment with antiseptics and sutur-
ating the site of enema tip pressure), multiple dilations of the agan-
glionic crypts.

A reliable sign of which is the presence of mobilized rectal
demucosation, dissected 2-4 cm below the mucocutaneous junction
(depending on the patient’s age), and fixed with four clamps.

After that, we continued to evacuate the colon to the marked
level of proposed resection. At the same time, the surgeon
of the team, who was working from the side of the abdomi-
cal cavity, carefully monitored the proper evacuation of
the colon.

We cut off all the layers of evagination step by step in a
transverse direction 0.5-1.0 cm from the dentate line and
applied sutures (PDS, vicryl 3/0-4/0) circularly between all
the layers of the colon and the rectal mucosa, thereby forming
colorectal anastomosis.

After applying anastomosis, we inserted a tube into the lu-
en of the evaginated colon above the line of the conjunction
to remove gases and feces in the postoperative period.

The rectal tube was fixed with a ligature to the skin of
the buttocks on the opposite side from the place of fixation of
the retrocolic drainage.

All the patients survived. In 15 (2.51%) out of 597 chil-
dren, in the early postoperative period, there were observed
surgical complications: retrocolic hematoma (n = 2), retro-
colic abscess (n = 7), anastomotic leak (n = 2), adhesive
intestinal obstruction (n = 3), intussusception (n = 1). Retro-
colic hematoma was conservatively corrected in both patients.
In case of retrocolic abscess and anastomotic leak, it was nec-
essary to impose a right-sided protective colostomy and to
proceed conservative treatment of the abscess and leakage. We
closed colostomy after 2-4 months in case of complete healing
of all complications and the absence of severe anastomosis
stenosis. Relaparotomy and viscerolysis were performed in
case of adhesive intestinal obstruction. Intussusception was
corrected by disinvagination in repeat laparotomy.

Fourteen (2.36%) patients developed surgical complica-
tions in the long-term period: residual aganglionosis (n =
9), anastomotic stenosis (n = 4) and coloposis. All children
with residual aganglionosis underwent sphincterotomy.
Colorectal anastomotic stenosis was corrected by bougienage
in three children. Repeated Soave-Boley surgery with colorec-
tal anastomosis was performed manually in another patient
at eliminate anastomotic stenosis. In coloposis, which was
the cause of recurrent constipation and abdominal pain, there
was performed relaparotomy with resection of the transverse
colon.

Repeated Soave-Boley operation with manual colorectal
anastomosis was successfully performed in 26 (4.362%) pa-
tients after primary correction of HD in other clinics using
different methods. The reasons for re-correction of HD were
as follows: residual aganglionosis (n = 1) after the Rehbein
procedure; residual aganglionosis (n = 2) and the presence of
a "sail" between the stumps of the rectum and colon at the site
of colorectal anastomosis (n = 2) after the Duhamel procedure;
residual aganglionosis (n = 2) after the classic Soave proce-
dure; residual aganglionosis (n = 10) and stenosis of colorectal
anastomosis (n = 6) after the Soave-Boley operation with man-
ual colorectal anastomosis; residual aganglionosis (n = 2) after
the Lynn procedure (sphincterectomy); anastomotic stenosis (n = 2) after laparoscopy-assisted TEPT. The technical capabilities and the results of the early and long-term periods allowed us to assert that the Soave-Boley technique with manual colorectal anastomosis is the operation of choice for secondary surgical correction of HD by any other means.

Periodic episodes of fecal smearing in the remote period in 45 (7.53%) children were eliminated by conservative treatment, which was carried out in specialized centers. Moreover, the psychological state and the persistence of parents and patients played a significant role in the adaptation period. The goal of HD treatment consisted in both medical and social rehabilitation of children, providing their early adaptation in society.

In general, the experience in treating HD with other methods of open surgical correction (the Swenson’s pull-through operation (n = 14), the Duhamel procedure (n = 29), the Rehbein method (n = 7) and the Soave procedure (n = 59)) helps conclude that the Soave-Boley procedure with manual colorectal anastomosis has significant technical advantages. The Soave-Boley technique allows better control over applying each suture and anatomical joining the edges of the rectum and colon. This reduces trauma to the rectal mucosa, contributes to the high reliability of colorectal anastomosis formation. The success of this technique is confirmed by a much lower number of early (2.51%) and late (2.36%) postoperative surgical complications, as compared to those after using other methods of open surgical correction - 17.52% and 16.35%, respectively.

**Conclusions**

1. The method of surgical correction of HD in children by the Soave-Boley technique with manual colorectal anastomosis allows better control over applying each suture, anatomical joining the edges of the bowel, and reducing trauma to the rectal mucosa, which ensures the high reliability of colorectal anastomosis.

2. According to the technical capabilities and the results of the early and long-term periods, the Soave-Boley technique with manual colorectal anastomosis is the operation of choice for secondary surgical correction of HD by any other methods.

**Conflict of Interest**

The authors declare that no conflicts exist.

**Financial Disclosure**

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**References**


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