Clinical Features of Early Diagnosis and a Choice of Method of Surgery for Hirschsprung Disease in Infants

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Abstract

The aim of this study is to improve the results of surgical treatment of Hirschsprung's disease in infants by choosing the most effective method.

This article presents the results of the treatment of Hirschsprung's disease in 61 patients of infancy. However, immediate and long-term results of the children's treatment with HD, operated on in infancy, using the traditional Soave-Boley method and modern minimally invasive technologies according to TENTC and the Georgeson method, which show different results, have been studied. Among 26 patients who had been operated on by the Soave - Boley method, the following results were obtained : excellent - in 10 (38.4%); good - in 8 (30.7%); satisfactory - in 6 (23%) and unsatisfactory - in 2 (7.6%). Among 29 patients operated on by the TEPT method, except for one patient, excellent and good results were obtained in 28 (96.6%) patients. What is more, among 6 patients who had been operated on by the Georgeson method, excellent and good results were obtained in all cases; there were no satisfactory or unsatisfactory results. It should be noted that among the operated HD patients according to the Soave-Boley method, there were severe patients, of which 7 patients had a colostomy in the neonatal period.

At the same time, radical one-stage surgical treatment for distal forms of HD using minimally invasive TEPT method is a priority in infancy, which can reduce complications to a minimum with improved functional outcomes of treatment, relative to two-stage treatment.

Keywords: *Hirschsprung's disease, treatment, surgical, diagnosis, pediatric.*

Introduction

Recently, the authors have recommended effective early diagnosis and surgical correction, the priority of which is a one-stage radical correction of Hirschsprung disease (HD) and the use of minimally invasive method. In this regard, the choice of optimal method for surgical treatment of HD in infancy is of particular relevance.

Regarding the reliability of the diagnosis of children with HD, taking a biopsy from the rectal mucosa is the «Gold Standard» for morphological diagnosis[1]. However, there is evidence of controversy and debatable questions about the validity of biopsies in young children with HD in literature[3]. Many authors claim that by the time of birth, intramural, ganglion cells still do not reach full morphological maturity[6]. They are especially immature in the caudal part of the colon, in the submucosal layer in particular[8]. This immaturity is physiological; final maturation occurs gradually. Consequently, it is believed that at an early age, a rectal biopsy is contraindicated and unreliable[22]. If this concept is not taken into account and take a biopsy in cases of suspected GB, then this diagnosis will be made often and unreasonably, especially in premature infants[23].

Other authors believe that the lack of preliminary morphological diagnosis and the preservation of the proximal dysganglionic segment of the colon during radical surgical treatment leads to unsatisfactory results[5].

At the present stage, the cases when there were no clear signs of HD according to irrigography became the indication[4] for carrying out a laparoscopic biopsy of the colon[21].

There is information that in some cases the histological structure of the colon also had a deviation from the norm. In 50% of children[9], a histochemical reaction suggested a diagnosis of intestinal agangliosis, in other cases, it only complicated the further diagnostic search[10]. So, low information content of histochemical diagnosis is explained by the absence of not only a nerve ganglia and nerve cells in the intestinal wall[12], but also parasympathetic fibers with their total lesion[13].

The available histochemical and morphohistochemical research method for determining the activity of tissue AChE in HD have been carried out for a long time with high reliability[2], however, its radical applications are not always available[11]. The modern neurohistochemical express method for a short research time provides information with high reliability (97-99%), however, it requires highly qualified specialists, expensive equipment and reagents that are not always available[18].

As well as the inclusion in the examination program of infants suffering from chronic constipation to identify HD, a morphohistochemical method[11], was not always reliable, which was not always reliable14, moreover, the applied clinical, radiological and functional method did not always make it possible to make a correct diagnosis[16]. Thus, many-sided and debatable questions on determining the localization of taking biopsy material from the rectal mucosa or laparoscopically from all the muscular - submucous layers of the colon and the low reliability of the biopsy in the diagnosis of HD remain open. Moreover, in an inaccessible situation, pediatric surgeons need to optimize and accurately solve the issues of intraoperative diagnosis, measures for the prevention of complications and relapses of the disease in infants.

Moreover, in surgical treatment of HD in infants, against the background of many concomitant diseases, malformations of other organs and morphofunctional immaturity of a child's body[7], preoperative preparation and postoperative management of patients causes particular difficulties[2]. On the other hand, the use of existing modified abdominoperineal proctoplastics (APR) using Soave technology (Boley, Lenyushkin) is more traumatic[17]. Surgeries of lowering the colon through the serous-muscular case of the rectum according to the Soave method with the creation of a "seamless anastomosis" in 2 stages are the safest for a patient, and their effectiveness reaches 90% in children older than infancy[12]. Fundamentally, the operation of laparoscopically assisted intestinal reduction is not different from the operation of Soave, however, it allows treatment in one stage and has all the advantages of minimally invasive interventions[13].

Regarding the results of surgical treatment of HD in children, according to the authors, in the postoperative period, having bowel movements is disturbed in 37% of sick children. At the same time, astriction occurs in about 8-20% of children, enuresis occurs in 5-26% of cases, which is unsatisfactory and leaves much to be desired[18]. The authors note that the introduction of minimally invasive method for the correction of Hirschsprung disease has led to a change in the spectrum of complications, the main of which were : colorectal anastomosis failure (3.4%) and its stenosis (8.5%)[22].

Therefore, in order to prevent postoperative complications and relapses of the disease in infancy, each method of operation according to Soave - Boley, Georgeson and Transanal endorectal pull-through (TEPT) according to Delatorre - Mandragon and Ortego - Salgado, in solving tactical and operational-technical issues requires optimization of indications, a differentiated choice of method depending on the anatomical form, degree of compensation and the course of the disease, which is an urgent problem in pediatric surgery.

Objective: Improve the results of surgical treatment of Hirschsprung disease in infants by choosing the most effective method.

Materials and Method

The immediate and long-term results of surgical treatment of HD in 61 infants were studied by us for the period 2010-2019. Patients' age: up to 3 months. - 8 (13.1%); 3-6 months - 17 (27.8%); 6-9 months - 20 (32.7%); 9-12 months - 16 (26.2%). By the forms of the disease were: rectal - 13 (21.3%); rectosigmoidal - 43 (70.4%); subtotal - 5 (8.1%). According to the clinical course: acute - 9 (14.7%); subacute - 39 (63.9%); chronic - 13 (21.4%). Congenital concomitant pathologies occurred: Down's disease - 1, aplasia and hypoplasia of kidneys - 3, coccyx agenesis - 3. There were 53 boys, 8 girls.

All children underwent general physical, laboratory, radiation (plan radiography, polypositionalirrigography, U/S) and morphological (macro preparation of colon agangliosis zones resected during radical surgery) studies.

Patients were divided into 2 groups: 1 - group (control) - 29 patients operated on by the Soave - Boley, Lenyushkin method; 2 - group (main) 32 operated on patients, among them: 26 cases by the TEPT method (Delatorre-Mandragon) and in 6 cases by the Georgeson method.

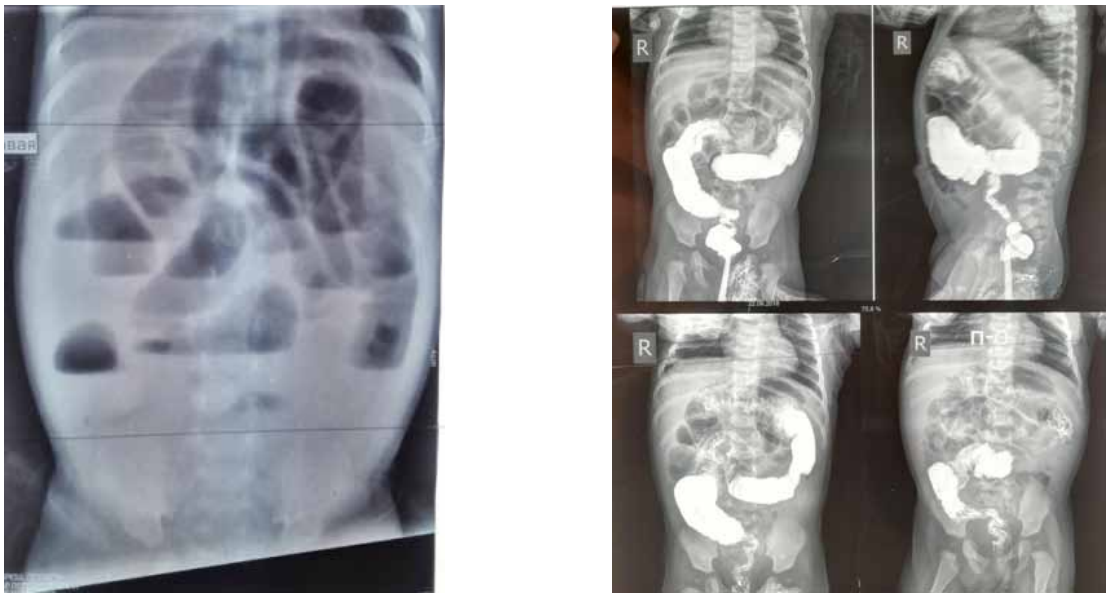
The effectiveness of each method has been studied by evaluating on a point scale according to the questionnaire data of operated patients in a remote period of 1-4 years. Clinical results were evaluated as: excellent, good, satisfactory and unsatisfactory.

Results and Discussion

Early diagnosis of HD in infants has its own specific features, and at the same time, the degree of maturity of the child, the age category, associated pathologies, the length of the aganglinal and the degree of (compensatory) expansion of the suprastenotic zone are of no small importance.

Enterocolitis is known to be a serious complication of HD in pre- and postoperative periods. Enterocolitis can develop at any age, from the neonatal period to the older age, regardless of medication or surgery. Recurrent enterocolitis can occur even in the presence of an externalized colostomy [].

According to our observations, during the neonatal period and early infancy (up to 3 months) there were 7 patients who received a clinical picture of acute low intestinal obstruction, a history of which noted: diarrhea, bloating, fever, anxiety due to spastic pain in abdomen, intoxication, hypovolemia and loose bowel movement with an admixture of blood. At the same time, the first thing was to exclude enterocolitis associated with Hirschsprung disease. After a plan radiography of the abdominal cavity, these patients underwent irrigography (Fig. 1a, b).



(A) **(B)**

Fig. 1. A - plan radiography of the abdominal cavity; B - Irrigography

In which the narrowed (aganglionic) zone in the distal colon was clearly contrasted, the transition zone in the form of a funnel-shaped expansion over the affected area in 5 patients with subtotal and 2 patients with a long zone of rectosigmoid form of the disease and manifestation of enterocolitis.

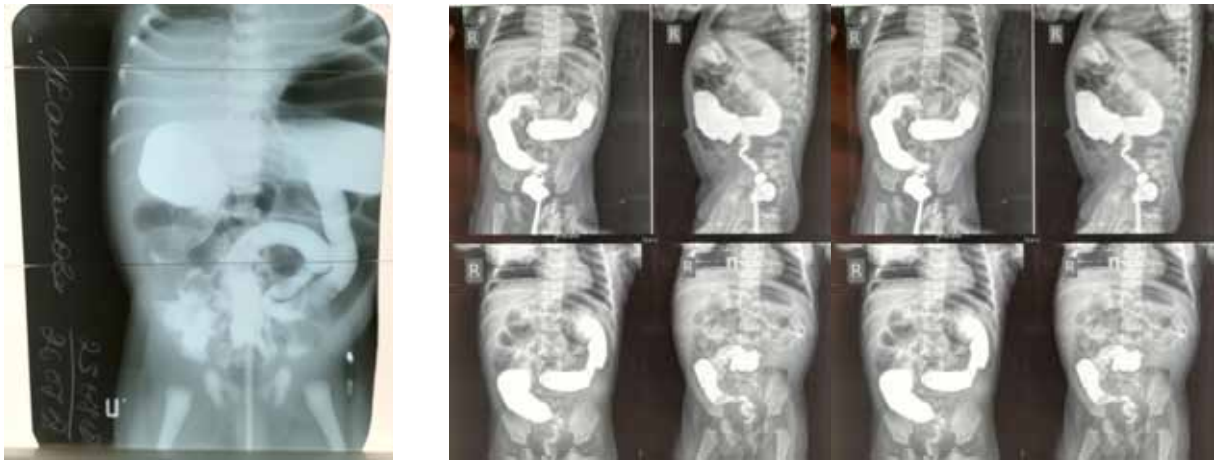
Having concomitant enterocolitis against the background of irritation of the intestine with spasmodic areas that was created with a false appearance of pathological narrowing. It means that during the period of newborn and early infancy, the risk of false-positive, and false-negative results of irrigography increases by

almost 3 times. Therefore, the x-ray area of narrowing does not always coincide with the area of the taken biopsy and agangliosis.

And also in 6 hypotrophic, functionally immature infants with enterocolitis in the initial early diagnosis of HD, the implementation of a layer-by-layer rectal biopsy on the inflamed mucosa and thinned intestinal wall was impossible.

That is why, in almost all cases, due to inaccuracy, inaccessibility and sometimes the lack of possibility of biopsy immunohistochemically, morphohistochemically and neurohistochemically, we did not carry out express method [11,14,15,19].

In patients (in 19 cases) older than 3 months with HD with a long aganglionic zone, a more pronounced picture of obstruction with high pressure had been observed with increasing suprastenotic expansion of the colon - left subtotal and long rectosigmoid form (Fig. 5a., b.).



(A) **(B)**

Fig. 2. Hirschsprung disease in infants: (A) left-side subtotal; (B) rectosigmoid form with a long zone of aganglionosis.

In a short zone with compensatory courses of the disease (in 40 cases), it is not always shown as in the classical form: narrowed, transitional zone with suprastenotic funnel-shaped extensions of the colon. At the same time, the sensitivity and specificity of irrigography in newborns and infants was significantly lower due to only breastfeeding.

With supplementary feeding and the transition to mixed, artificial and natural feeding, the clinical picture of chronic bowel movement retention began to increase in patients, and against this background, the use of polypositionalirrigography in these patients has greatly expanded the range of diagnostic capabilities of HD in infants.

Despite the limitations or sometimes difficulties of using morphological and other research method in the early diagnosis of HD in infants, we consider polypositionalirrigography and preoperative diagnostic laparoscopy to be more optimal and reliable method. We carried out preoperative diagnostic laparoscopy with dubious diagnosis of HD and the length of the zone of aganliosis, taking into account the choice of the method of operation in 6 patients.

In the surgical treatment of HD, until recently, though STI according to Soave - Boley and Lenyushkina, which we performed in the first group of patients in 25 cases, were considered optimal and more physiologically substantiated method in children. These operations are performed in 2 stages, the abdominal stage is almost identical, but they are not free from shortcomings and complications. At the abdominal stage, in the depths of the pelvis against the background of technical difficulty during demucosis of the rectal wall, as close as possible to the anus, internal anal sphincters are often damaged and perforation occurs, which often causes pelvic peritonitis, interfacial abscess, and stenosis.

Moreover, with excessive blind demucosis of the intestine, blunt trauma (with instruments) and impaired blood and lymph formation in the adjacent pelvic organs (sphincter muscles, ovaries, prostate gland, urethra) in the long term, the consequences often lead to fecal and urinary incontinence, as well as are the background state in the development of diseases of the reproductive organs in boys and girls.

However, at the modern level, with the improvement of new minimally invasive and high-tech TEPT operations with video-assisting at the abdominal stage - with Georgeson's method and without it, we had the opportunity to operate sick children with HD during the neonatal and infancy years.

With the gain in experience, we have developed various tactical approaches for choosing the method of operation and optimizing the indications depending on the anatomical shape, degree of compensation, and the course of HD.

In case of subtotal and proximal left-sided long forms of HD disease in 6 cases (agangliosis zone length over 30 cm), in the main group of patients, in order to reduce intra-abdominal complications, the Soave-Boley method was replaced by laparoscopic diagnostics and colon mobilization. The perineal stage is an oncoming transanaldemucosis "from the bottom up" with the reduction of the mobilized intestine. After resection of the agangliosis zone, a primary colorectal anastomosis (Georgeson operation) was created, the results of which were successful outcomes.

In rectal and rectosigmoid forms of HD in infancy, a long zone of agangliosis in the rectum and sigmoid colon (up to 30 cm) was performed by TEPT (Delatorre - Mandragon and Ortego - Malgado), in which demucosis was performed by transanal access with lowering of the colon endorectally. After resection of the agangliosis zone, the primary colorectal anastomosis is transanally superimposed (Fig. 3.). The operation by using TEPT method was performed in 26 children with HD in infancy, a rectal and rectasigmoid form of the disease. Moreover, all operations were performed without prior colostomy.

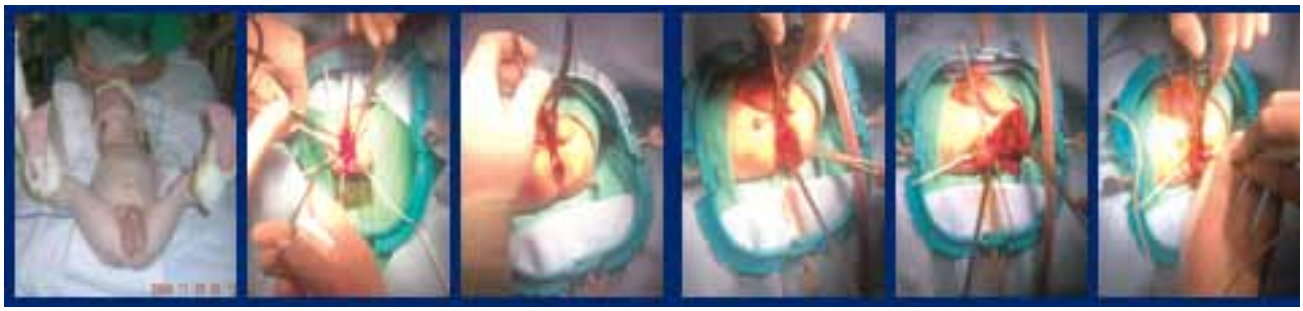


Fig. 3. TEPT - Delatorre - Mandragon and Ortego - Malgado

When performing TEPT, it should be noted that it is possible to determine the level with wide gut resection in order to prevent relapse of the disease, since the younger the age of patients, the less clearly and contrast the narrowed area and suprastenotic enlarged part of the intestine are manifested during irrigography. This is most characteristic of subacute (rectal and rectasigmoidal form) and atypical (enterocolitis associated form) clinical course of the disease, which complicates early diagnosis and intraoperative determination of the level of resection. For that purpose, to determine intraoperatively the location of the level of wide resection and ensure the optimal radicality of TEPT method, we studied morphologically the state of the histostructure of the nerve plexuses, nodes, cells and fibers in the wall of the transitional and enlarged parts of the removed intestine near the gross specimen. At the same time, in 9 patients with HD, the gross specimen obtained by wide resection of the colon was subjected to a morphological study, the reason for which was the additional loops with dolichosigmoid.

The initial, middle and upper parts of the suprastenotic enlarged zone of the colon (Fig. 4). has been studied morphologically.

Fig. 4. A gross specimen obtained by a wide resection of the colon.

At the same time, in the initial section of the enlarged part of the intestine, at a distance of 3 cm from the transition zone, areas of dysgangliosis were noted, characterized by places of absence, atrophy, destruction, hypertrophy and hyperplasia of the preserved nerve plexus nodes.

Further, when studying the mucous membrane of the middle suprastenotic enlarged part of the intestine, at a distance of up to 5 cm, from the transition zone, their atrophicity was noted, expressed in wide gaps in the crypts, diffuse nature of the edema, loosening of its own connective tissue plate with foci of inflammatory infiltrate and sclerosis. There are relatively hypertrophic and hyperplastic nerve nodes due to swelling of nerve cells, fibers, and Schwann's cells hyperplasia (Fig. 5a).

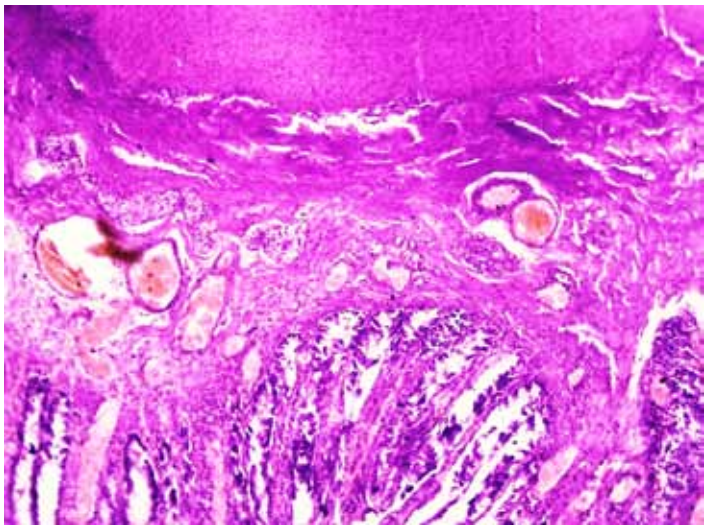


Fig. 5a. The submucous layer of the suprasthenic part of the intestine with HD in an infant is up to 3-5 cm long . Hypertrophy and hyperplasia of the nerve plexuses against the background of chronic inflammation. H & E stain Magn: 10x20.

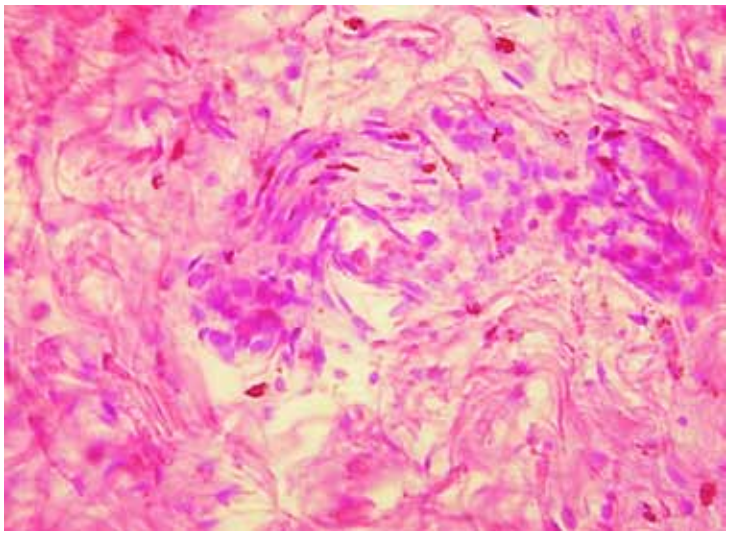


Fig. 5b. The submucous layer of the suprasthenic part of the intestine with HD in an infant is up to 3-5 cm long . Hypertrophy and hyperchromasia of Meissner nerve nodes with hyperplasia of Schwann's cells. H & E stain Magn: 10x40.

Nerve cells are located in one pole of the node with signs of hypertrophy and hyperchromasia, in the other pole of the node there are nerve fibers from hyperplastic Schwann's cells (Fig. 5b). In the submucosal layer of the middle expanded part of the intestine, at a distance of up to 5 cm from the transition zone, the presence of nerve nodes of various size and composition is noted. In these nerve nodes there are few nerve fibers and glial cells and they are located on the periphery.

Meissner nerve nodes of round shape with a small number of nerve cells (Fig. 6a), where, on the contrary, there are more nerve fibers circles of Schwann's cells form a thin membrane (Fig. 6b), which covers, depending on the degree of decompensation. These secondary morphological changes at the level of the less pronounced dilated "normal" part of the intestine lead to normalization of the structure of nerve nodes in the mucous and submucosal layer up to 5 cm from the transition zone, depending on the degree of compensation of the suprastenotic expansion of the colon.

12.30 ± 0.68
9.80 ± 0.55

Fig. 6a. The submucous layer of the suprasthenic part of the intestine with HD in an infant is up to 3-5 cm long . Meissner nerve nodes with a large number of nerve cells and a thin myelin sheath. Magn: 10x40.

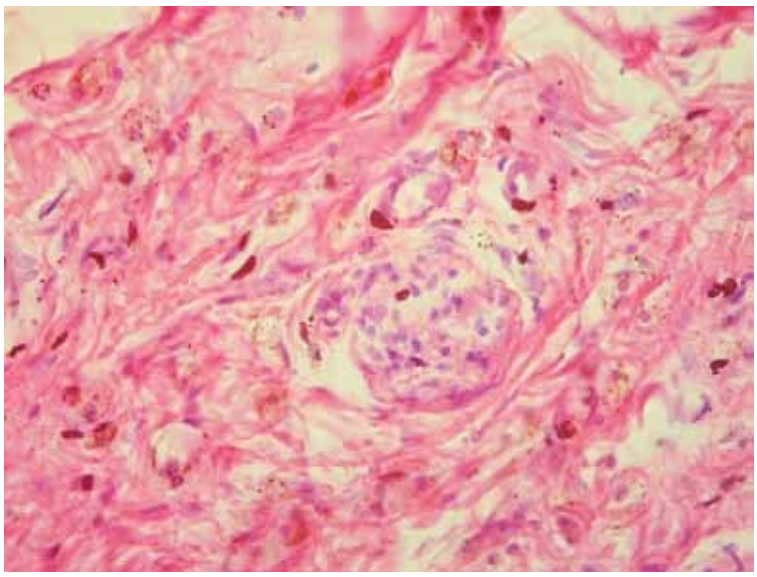


Fig. 6b. The submucous layer of the suprasthenic part of the intestine with HD in an infant is up to 3-5 cm long. . Meissner nerve nodes with a small number of nerve cells and a large amount of nerve fibers. H & E stain Magn: 10x40.

In the intramuscular layer of the enlarged part of the intestinal wall, separate Auerbach nerve nodes with dystrophic, atrophic, and destructive changes up to 5 cm long are noted (Fig. 7a). In this case, mainly nerve cells, glial cells and nerve fibers underwent dystrophic changes, and Schwann's membranes underwent atrophic changes. Microscopic examination of the wall of the intermediate part of the expansion of the colon shows a picture of the proliferatively inflammatory form of chronic colitis. The muscle layer is thickened due to hypertrophy of smooth muscle cells and the presence of interstitial lymphohistiocytic infiltrate with atrophy and destruction of the nerve ganglia. Auerbach's nerve nodes are compressed by inflammatory infiltrate, atrophic nerve cells are compressed and small, hyperchromatic. Pathomorphological changes of the nervous system of the colon correspond to hypogangliosis (Fig. 7b).

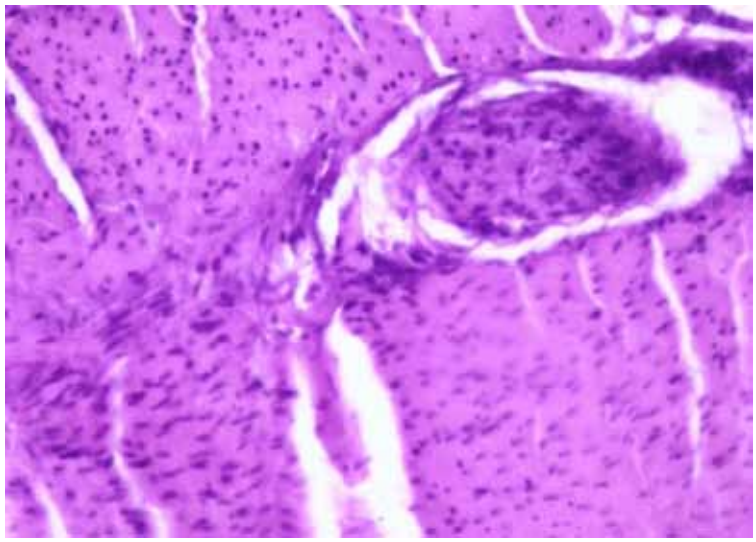


Fig. 7a. The muscle layer of the suprasthenic part of the intestine is up to 3-5 cm long. with HD in an infant. Separate Auerbach nerve nodes with dystrophic, atrophic and destructive changes. H & E stain Magn: 10x40.

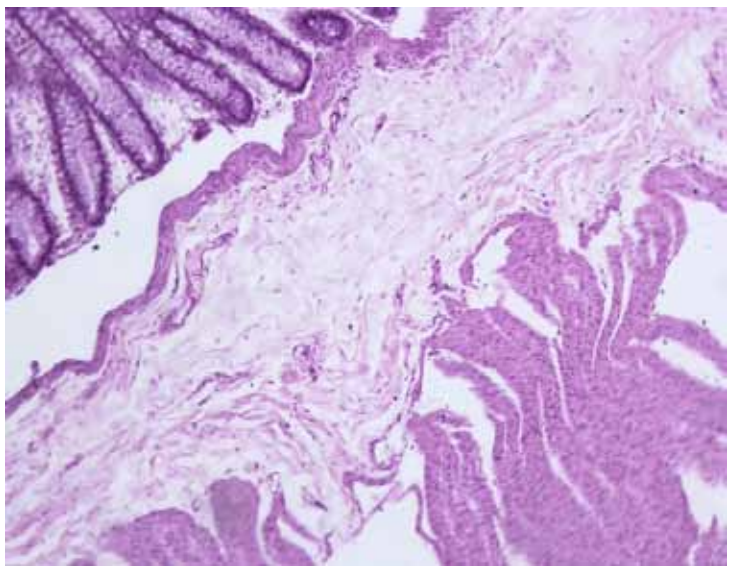


Fig. 7b. The muscle layer of the suprasthenic enlarged part of the intestine up to 5 cm long. Auerbach's nerve nodes are compressed by inflammatory infiltrate, atrophic nerve cells are compressed and small, hyperchromatic. The Auerbach nerve plexuses correspond to colon hypogangliosis. H & E stain Magn: 10x40.

Moreover, in different topographic sections of the wall of the enlarged part of the intestine, the Auerbach nerve plexuses differ in histoarchitectonics and sizes from each other. In the first section, where the mesentery is attached there are neurovascular bundles and nerve nodes are larger, there are many cells located rarely in one microscope objective, there is only one node. The farther the section of the intestinal wall from the zone of attachment of the mesentery, the denser the Auerbach nerve plexuses are located, in one microscope objective there are 2-3 nodes. They have few nerve cells, the nerve fibers are loosened, and the circumferences of the myelin sheaths are thick due to hyperplasia of Schwann's cells.

Thus, at the level of the suprastenotic enlarged zone at a distance of up to 5 cm, thesigns of irreversible sclerosis and fibrous degeneration of the muscle layer, degenerative -inflammatory changes in the mucous membrane,dysganglionic "inferior" sections of the nerve plexuses, which can subsequently serve as a source of development of hypoperistaltics, the cause of chronic colostasis in the long term after surgery have been diagnosed. Based on the data of histomorphology, we can conclude that to determine the optimal level of the upper border of the resection for rectal and rectosigmodal forms of HD, even with minor dilatation of the suprastenic zone, there are varying degrees of dysganglionic and "inferior" sections of the nerve plexuses. This is why, a wide resection at a distance of at least 5 cm from the enlarged zone is recommended and it is considered to be an optimal site for the resection zone and prevent recurrence of the disease.

Such intraoperative tactics for determining the level of wide resection in the mobilized part of the intestine was used in 28 patients with TEPT surgery, and in 8 patients it was established with diagnostic laparatomy. In the remote period, all patients with no signs of chronic constipation obtained excellent and good results.

At the same time, we have proved that the place and level of wide resection of the suprastenotic zone in infants depending on its compensatory expansion for the prevention of postoperative chronic constipation and relapse of the disease in a more distant period after surgery is of natural significance.

The advantages of TEPT were: radicalism; low injuries; one-stage; the absence of postoperative complications from the abdominal cavity and pelvic organs; smooth postoperative course. Contraindications were: severe vital malformations, acute severe enterocolitis, severe dilatation of the proximal intestine, severe general somatic state of the child, Hirschsprung associated enterocolitis (HAE), multiple, deep erosive ulcers in the mucosa of the rectum ampule and purulent inflammation of the parianal region.

In solving the tactical issues of diagnosis and treatment of HD in infancy, enterocolitis is the main and prognostically key condition for postoperative complications and mortality. Upon admission to the intensive care unit, newborns with HAE undergo intensive therapy, which includes correction of water-electrolyte and protein balance, cleansing and therapeutic enemas. If the length and localization of the agangliosis zone with an acute course of the disease is inaccurate, a colostomy is first applied above the site of obstruction and a floor biopsy from the distal sections is simultaneously performed. Further, when agangliosis is detected histologically, the zone in the lower part of the ostomy, within 3-6 months after the complete elimination of enterocolitis, the second stage is the operation of Soave-Boley with the simultaneous elimination of colostomy. We used this tactical approach in 3 patients with enterocolitis associated with HD, in whom, after surgical treatment, a successful fate was obtained in the late late period.

Also, we conducted 6 newborns and young children who had been accepted from the region with a severe course of HAE, where against the background of diarrhea syndrome, severe dilatation of the proximal intestine and with a general somatic status, a colostomy was imposed on an emergency basis. In the future, after removing the patient from the septic state and treating enterocolitis at the age of 7-9 months, a radical Soave-Boley operation was performed in the second stage with the simultaneous elimination of colostomy.

In other cases, with an uncomplicated form of HD and with the ex ception of total agangliosis, a one-stage correction of HD was performed. With incomplete primary diagnosis and with the presence of a very long zone of agangliosis, before TEPT, we used laparoscopic diagnostics in 10 patients to clarify the areas of elongation and expansion, localization and the true length of the zone of narrowing and suprastenotic expansion. After clarification of the above, the optimal location of the resection level was determined, the marks were left on the suprastenotic enlarged area, 5 cm from the transition zone, the proximal part of the colon agangliosis was

mobilized by separation of adhesions and the fixative ligament, where conditions for the possibility of TEPT were created.

With subtotal, total and left-sided disease with a long zone of agangliosis, clipping and resection of the blood supplying large mesenteric vessels of the second and third order. Then, a pull-through operation of the large intestine (Georgeson's operation) through the proctal access was performed . Moreover, the crucial points are: a meticulous execution technique, adequate hemostasis, maintaining good blood supply for the prevention of ischemia and preventing torsion and tension of the reduced intestine. These points prevent possible com plications in the postoperative period of HD in young children.

In acute type of HD, in the presence of intestinal obstruction, HAE with toxic dilatation of the colon, perforation of the blind and other parts of the colon, was carried out by colostrum with biopsy at different levels of the colon and rectal biopsy. With the subtotal and total form of HD in 3 cases, a Soave - Boley operation was performed with the simultaneous elimination of the colostomy. With the subtotal and total form of HD in 3 cases, the Soave-Boley operation was performed with the simultaneous elimination of colostomy.

According to our clinical observations, in all cases, complications in the intra- and postoperative period associated with the technique, the operation was not observed. In the postoperative period, in all cases, a Foley catheter No. 28-32 was inserted into the rectum, and turunda soaked in «Bakstims» balsam was left between the coloanal anastomosis and periodically changed after 2 days. The course of the postoperative period in all patients proceeded satisfactorily. Patients were usually discharged from the hospital 7 days after surgery.

Enterocolitis occurred in one patient in the early postoperative period. Stenosis in the area of the anastomosis often caused partial low intestinal obstruction against the background of circular and segmental-circular anastomosis, and was not a high risk factor. At the same time, the passage of a smaller Foley catheter through the anastomosis, the picture of mechanical intestinal obstruction was quickly eliminated. Two weeks later, during the control finger test, part of the stenosis was eliminated conservatively by dilating the narrowed area and rupturing the absorbable sutures of the colorectal anastomosis, which was carried out easily.

In the postoperative period, the failure of the coloanal anastomosis in our observations has never occurred. We didn't come across violations of bowel movement, including constipation, incontinence and enuresis. Functional constipation can be treated with conservative method, such as cleansing enemas and diet.

In addition to the use of Georgeson's surgery and TEPT, a 3-fold decrease in the number of postoperative complications was obtained in comparison with the control group.

During the monthly follow-up examination, complaints of constipation of varying intensity were observed only in 35% of patients. In these patients, with a long period, there were clinical signs of constipation until the filament was completely absorbed (up to 3 months) at the site of the anastomosis, due to its partial narrowing. At the same time, cleansing enemas and bougienage of stenosis to the age norm were performed. In order to restore the neuro-reflex connection in the anorectal zone, 1-3 courses of electrical stimulation were performed. In this case, the signs of functional disorders of the stool gradually disappeared and the normal act of defecation was restored.

To study long-term results, questionnaires were conducted 3 months after the radical operation, in 45 sick children with HD at the age of 1-3 years, by scoring on a point scale. The list of questionnaire questions is presented in table No. 1, where clinical violations of the act of defecation were evaluated as: good, satisfactory, unsatisfactory.

Table 1: Questionnaires to assess the long-term results of surgical treatment of Hirschsprung disease.

Symptoms	Long-term results					
	Good	Point	Satisfactory	Point	Unsatisfactory	Point
1. Abdominal distention	No	10	Often	5	Constantly	0
2. Difficulty in the act of defecation	No	10	After 3 days with difficulty, but independently	5	After 4-6 days, not on their own, after an enema	0
3. Incomplete bowel movements	No	10	Partially	5	Yes	0
4. Reducing bowel movement diameter	No	10	Partially reduced	5	Sharply reduced	0
5. Lack of a bowel movement and the urge to defecate	No	10	Partially	5	Yes	0
6. Fecal incontinence without the urge to defecate	No	10	Partially	5	Yes	0

Moreover, if the result was 45-60 points, we rated it good; 30-45 points - satisfactory; below 30 points - unsatisfactory.

Long-term results of surgical treatment of Hirschsprung disease in infants by choosing a method are presented in Table 2.

Table 2: Long-term results of surgical treatment of Hirschsprung disease in infants by method choices

Anatomical forms	Method operations	Amount of operated Patients (%)	Long-term results (n- 61)			
			Unsatisfactorily	Satisfactorily	Good	Excellent
Rectal 13(21.3%)	TEPT	13(21.3%)	-	-	34.9%)	10(16.4%)
Rectosigmoid 22 (36.1%)	TEPT	10(16.4%)	-	-	2(3.2%)	8(13.1%)
	Soave - Boley	12(19,7%)	-	2(3.2%)	5(8,1%)	5(8,1%)
Rectosigmoid with a longzone19 (31,1%)	TEPT	6(9.8%)	-	1(1.6%)	2(3.2%)	3(4.9%)
	Soave - Boley	7(11.5%)	-	1(1.6%)	1(1.6%)	5(8,1%)
	Georgeson	6(9.8%)	-	-	2(3.2%)	4(6.5%)
Rectosigmoid with a long zone + HAE2(3,2%)	Soave - Boley	2(3.2%)	-	1(1.6%)	1(1.6%)	-
Subtotal form + HAE + colostomy 5 (8.1%)	Soave - Boley	5(8,1%)	2(3.2%)	2(3.2%)	1(1.6%)	-
Total		**61(100%)**	**2(3,2%)**	**7(11,5%)**	**17(27,9%)**	**35(57,3%)**

At the same time, we obtained long-term results in general for surgical treatment of HD in infancy: excellent - 57.3%, good - 27.9%, satisfactory - 11.5%, unsatisfactory - 3.2%.

Among 26 patients who had been operated on by the Soave - Boley method, the following results were obtained : excellent - in 10 (38.4%); good - in 8 (30.7%); satisfactory - in 6 (23%) and unsatisfactory - in 2 (7.6%). Among 29 patients operated on by the TEPT method, except for one patient, excellent and good results were obtained in 28 (96.6%) patients. And also among 6 patients operated according to the Georgeson method, excellent and good results were obtained in 100% of cases, there were no satisfactory and unsatisfactory results. It should be noted that among the operated HD patients according to the Soave-Boley method, there were severe patients, of which 7 patients had a colostomy in the neonatal period.

Thus, according to the study of the long-term results of the first group of patients operated on by the Soave-

Boley method: in the late period there was stenosis in the anastomotic zone with a violation of the bowel movement as a constipation with complete and partial absence of a feeling of urge. For children with satisfactory and unsatisfactory results, a stationary examination was proposed with irrigography, ultrasound of the abdomen and perineum, if necessary, examination of feces for coprology and dysbacteriosis, rectal ulceration, ESM and the complex rehabilitation treatment post-mortem until the symptoms disappear for 1-3 years.

In the second group of patients operated on by the TEPT method in the remote period after constipation surgery, they were episodic in nature, and urinary incontinence was not observed. Fecal incontinence was noted only in the form of a temporary nature by the type of increased stool and fecal incontinence were in the presence of loose stool. After receiving the ESM and bougieurage and the complex rehabilitation treatment, they gradually disappeared within 6–9 months.

Thus, the study of immediate and long-term results of the children's treatment with HD, operated on in infancy, using the traditional Soave-Boley method and modern minimally invasive technologies according to TENTC and the Georgeson method, shows different results.

At the same time, radical one-stage surgical treatment for distal forms of HD using minimally invasive TEPT method is a priority in infancy, which can reduce complications to a minimum with improved functional outcomes of treatment, relative to two-stage treatment.

Results

1. In the surgical treatment of distal forms of HD in infants, the TEPT method is the method that is preferred for selection and the colostomy is not placed on.
2. With proximal and decompensated forms of HD with the presence of a colostomy, the optimal method of radical surgery is abdominoperineal proctoplastics according to the Soave (Boley and Lenyushkin) technology with the simultaneous elimination of the colostomy.
3. In the complicated, acute course of the subtotal form of HD during the neonatal period, primary emergency colostomy application saves the life of a child and contributes to the treatment of bacterial and HAE and the subsequent use of abdominal-perineal proctoplasty according to the Soave technology (Boley and Lenyushkin) with the simultaneous elimination of the colostomy.

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