Original Article

Soave's transanal endorectal pull-through procedure for treatment children with Hirschsprung's disease

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Abstract

Procedeul endorectal tansanal Soave în tratamentul copiilor cu boala Hirschprung

Boala Hirschsprung (BH) are la bază malformațiile elementelor peretelui intestinal gros distal, a cărei esență este absența completă a ganglionilor neuronali intramurali. De-a lungul deceniilor au fost descries diferite tehnici chirurgicale de corecție a acestei maladii, actualmente cea mai răspândită abordare fiind procedeul endorectal transanal Soave (PETS).
Scopul lucării a fost în prezentarea propriei experiențe în managementul BH la copiii utilizând PETS cu analiza rezultatelor obținute.
Material și metode. În perioada 2014-2018, în clinica noastră au fost operați 15 pacienți cu BH utilizând PETS. Diagnosticul BH s-a bazat pe studierea atentă a anamnezei, metodele de examinare clinică obișnuite, inspecție manuală rectală, proctosigmoidoscopie, irigografie cu bariu, biopsia mucoasei intestinului gros cu examenul histologic și histochimic (activitatea acetilcolinesterazei), studierea microflorei intestinului gros.
Rezultate. De obicei, semnele și simptomele apar la scurt timp după naștere, dar uneori pot apăre și în perioade mai tardive. Constipațiile, meteorismul și vărsăturile au fost cele mai frecvente manifestări ale BH la copii. Clisma de contrast, care nu este o metodă specifică în diagnosticul BH, a fost efectuată în toate cazurile. Biopsia rectală cu examenul histopatologic au permis de a confirma diagnosticul. La toți cei 15 pacienți a fost constatat segmentul aganglionic de lungime standard - majoritatea pacienților prezentând segment de aganglioză localizat în regiunea recto-sigmoidă (66, 67%). În aceste cazuri a fost preferat procedeul Soave, realizat cu ajutorul tehnicii transanale într-o singură etapă la 11 (73,33%) pacienți și asistată laparoscopic într-o singură etapă la 4 (26,67%) pacienți. PETS asistată laparoscopic într-o etapă fără incizie abdominală a fost posibilă la pacienții cu BH cu prezentare tardivă și cu fecalom.
Concluzii:

1. PETS într-o singură etapă este tehnica chirurgicală sigură și eficientă în tratamentul pacienților cu BH cu afectarea segmentului rectal și rectosigmoid, rezultatele clinice fiind satisfăcătoare.
2. PETS asistat laparoscopic într-o singură etapă fără incizie abdominală a fost posibilă la pacienții cu BH diagnosticată tardive și cu fecalom.
3. PETS are principii anatomice optime pentru reconstrucția rectală în comparație cu alte tehnici chirurgicale utilizate în tratamentul BH la copii, fiind intervenția chirurgicală de elecție în patologia dată.
4. Complexul de examinare elaborate al pacienților cu BH permite nu numai un diagnostic precis al patologiei date, dar și depistarea precisă a cauzelor complicațiilor și tulburărilor funcționale intestinale în perioada postoperatorie.
5. Chiar și corectarea chirurgicală fără erori a BH la copii nu garantează succesul absolut fără un tratament de recuperare și reabilitare în perioada postoperatorie timpurie și târzie.

Cuvinte cheie: boala Hirschprung, diagnostic, tratament minim invaziv,

Abstract

Hirschsprung's disease is based on malformation of the distal large intestine wall's elements, which essence is either complete absence of intramural neural ganglia. Various surgical techniques have been described and employed over decades for the treatment of HD. The most popular approach for the children with HD today is the Soave's transanal endorectal pull – through (TEPT). *Aim* – to present own experience in the management of the HD in children with Soave's TETP method with the analysis of treatment results.

Material/methods. 15 patients were operated for HD with Soave's TEPT in our clinic between 2014-2018. HD diagnostics were based on careful studying of anamnesis, common clinical examination methods, manual rectal inspection, proctosigmoidoscopy, irrigography with barium, biopsy of a large intestine's mucous membrane for definition of histological and histochemical study (acetylcholineesterase activity), studying of large intestine's microflora.

Results. Usually signs and symptoms appear shortly after birth, but sometimes they're not apparent until later in life. Constipations, meteorism and vomiting were the most common sings of HD in children. Contrast enema is not a specific method for diagnosing diagnosis of HD was done in all the HD. Rectal biopsy and histopathology examination confirmed the patients. All 15 patient's hade standard length aganglionic segment – the majority of patients had segment of aganglionosis localized in the recto- sigmoid region (66, 67%). In cases used the prefered type of PT is the Soave approach, which is performed with the one – stage transanal technique by 11 (73,33%) patients and with one – stage laparoscopy - assisted by 4 (26,67%). One-stage laparoscopy – assisted TEPT by Soave without abdominal incision was feasible in patients with late-presenting HD and with fecaloma.

Conclusions.

1. One – stage Soave's TEPT is the safe and effective surgical technique for patients with rectal and rectosigmoid segment HD. The clinical outcome is satisfactory.
2. One-stage laparoscopy – assisted TEPT by Soave without abdominal incision was feasible in patients with late-presenting HD and with fecaloma.
3. Operation of endorectal pull-through by Soave method has optimum anatomic principles of a rectum reconstruction as compared to the other surgical techniques for the treatment of the HD in children and it is the operation of choice at the given pathology.
4. The developed examination complex of patients suffering from the HD allows not only to diagnose the abnormality itself, but also to reveal the reasons for complications and functional disorders of the brought down gut in the postoperative period in case of each child with big accuracy.
5. Even infallible surgical correction of HD in children does not guarantee absolute success in treatment without careful postoperative regenerative treatment and rehabilitation in the early and late postoperative period.

Keywords: Hirschprung disease, diagnosis, minimally invasive treatment

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Introduction

Hirschprung's disease (HD) is a condition that affects the large intestine (colon) and causes problems with passing stool. According to the modern concept based on clinical, functional, radiological and morphological data as well as on other research evaluations, Hirschsprung's disease is based on malformation of the distal large intestine wall's elements, which essence is either complete absence of intramural neural ganglia. The first description of this pathology dates back to 1886, when a Danish pediatrician Harald Hirschsprung presented the first portrayal of congenital megacolon in the Society of Pediatrics in Berlin [2]. Since then an abundant literature has given many controversial aspects of Hirschsprung's disease (HD) management. Various surgical techniques have been described and employed over decades for the treatment of HD, but all stem from the approaches described by Orvar Swenson in 1948 [5], Bernard Duhamel in 1956 [1] and Franco Soave in 1963 [4]. The most popular approach for the children with HD today is the Soave's transanal endorectal pull – through (TEPT).

The aim of this article – to present own experience in the management of the HD in children with Soave's TETP method with the analysis of treatment results.

Material and methods.

15 patients were operated for HD with Soave's TEPT in our clinic between 2014-2018. The age of patients was: beneath 1-year-old – 8 children (53,33%), 1-3 –years old – 4 children (26,67%), 4-5 years-old – 3 children (20%). HD is usually diagnosed in patients who are younger than 1 year, but in some individuals, it is found later, than 1 year old. HD diagnostics were based on careful studying of anamnesis, common clinical examination methods, manual rectal inspection, proctosigmoidoscopy, irrigography with barium, biopsy of a large intestine's mucous membrane for definition of histological and histochemical study (acetylcholine-esterase activity), studying of large intestine's microflora.

The majority of patients had rectosigmoid form of HD (10; 66,67%), rectal form of HD – in 5 children (33, 33%). Clinical outcome was assessed by interviews and questionnaires.

Table1. **Dynamics of HD clinical presentations**

Clinical presentations	Absolute number of patients	Percentage (%)
I. Early symptoms:		
- constipation with fecal impaction	15	100
- meteorism		
- anxiety	15	100
	11	73,33
II. Late symptoms:		
- anemia	11	73,33
- hypotrophy	11	73,33
- fecal bolus	4	26,67
- diarrhea	5	33,33
- trouble gaining weight	7	46,67
- growth delay	7	46,67
III. Complication symptoms:		
- vomiting	12	80
- abdominal pain	8	53,33
- malnutrition	11	73,33

Results and discussion

Dynamics of the clinical symptoms of HD in our patient's group has been presented in table 1. Usually signs and symptoms appear shortly after birth, but sometimes they're not apparent until later in life. Constipations, meteorism and vomiting were the most common sings of HD in children. A rectal examination may reveal a loss of muscle tone in the rectal muscles. This diagnostic clinical method we apply to all our patients. Contrast enema in patients with suspected HD is sill very popular, but it is not a specific method for diagnosing of HD was done in all the HD.

Rectal biopsy and histopathology examination confirmed the patients. We take the most distal biopsy at 2 cm or 3 cm from the dentate and used for HD diagnosis hematoxylin/eosin and acetylcholinesterase. Rectal biopsies are obtained using the open full-thickness in all patients (100%). To receive biopsy report it takes more than 3 days for all cases.

The timing of the pull – through operation is varies. We consider that the optimum time to perform a radical operation is the age of 3 – 6 months, which as we claim is due to:

- reduction of indications for prolonged preoperative preparation
- absence of the expressed secondary local and general character changes
- improvement of functional treatment results.

While waiting for surgery the bowel is maintained decompressed with rectal irrigation by 70% patients or with rectal dilatation by 30% children.

All patient's hade standard length aganglionic segment – the majority of patients had segment of aganglionosis localized in the recto- sigmoid region (66, 67%). In cases used the prefered type of PT is the Soave approach, which is performed with the one – stage transanal technique by 11 (73,33%) patients and with one – stage laparoscopy - assisted by 4 (26,67%).

The main feature of this operation is endorectal bringing down of a large intestine through demucosated rectal cylinder with a primary colo-rectal anastomosing at 1-1,5cm from anus. Endorectal bringing down with primary end -to – end anastomosis favorably differs when carried out in one stage, it is accessible to be performed in children in early age, simple in postoperative care and promotes early patient's activity. One-stage laparoscopy – assisted TEPT by Soave without abdominal incision was feasible in patients with late-presenting HD and with fecaloma. Rectal irrigation under general anesthesia and the use of laparoscopy and bipolar coagulator help to overcome the technical difficulties of his procedure.

There were no intraoperative or early postoperative complications. Patients started a diet a median of 3 days after the operation and were discharged a median of 12 days. All patients were followed up from 6 to 24 months. Clinical outcome was assessed by standardized interviews and questionnaires. During the first 6 months after Soave operation full normalization of function of the large bowel's reduced segment has occurred in 12 children which was 80% of the total. In children the mean stool times were 1 to 2 per day. Endorectal pull-through by Soave method has less impact on the sphincters. In 2

children large bowel's function disorders (grade I soiling) were still observed throughout the time of 2 years after operative intervention and they normalized after several courses of conservative regenerative treatment and rehabilitation had been carried out. 1 patient had stenosis of colo-rectal anastomosis. Colo-rectal stenosis anastomosis was subject for rectal dilatation in this patient.

Such postoperative complications as anastomotic leak, recurrence of constipation, recurrent enterocolitis have not been observed in our patients after SPTP.

Principal causes for coprogreasing (coproozing) are abnormalities of conditioned – reflex connections between the formed rectum and anus and also discoordination of the function of anal sphincters with reflex loss on defecation and increase of the endorectal pressure. In the presented group of the children correction of functional postoperative abnormalities was carried out by conservative treatment which included reflexotherapy, dietotherapy, exercise therapy, physiotherapeutic procedures, general and local pharmacotherapy with positive clinical effect. The organic reason for complication such as residual aganglionosis segment were subject for surgical elimination. Lynn's sphinctermyectomy was applied in this case where short aganglionotic areas – up to 5 cm long – was left [3].

Conclusions:

1. One – stage Soave's TEPT is the safe and effective surgical technique for patients with rectal and rectosigmoid segment HD. The clinical outcome is satisfactory.
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3. Operation of endorectal pull-through by Soave method has optimum anatomic principles of a rectum reconstruction as compared to the other surgical techniques for the treatment of the HD in children and it is the operation of choice at the given pathology.
4. The developed examination complex of patients suffering from the HD allows not only to diagnose the abnormality itself, but also to reveal the reasons for complications and functional disorders of the brought down gut in the postoperative period in case of each child with big accuracy.
5. Even infallible surgical correction of HD in children does not guarantee absolute success in treatment without careful postoperative regenerative treatment and rehabilitation in the early and late postoperative period.

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Conflicts of interest: *authors have no conflict of interest to declare*