Numerous surgical techniques for Hirschprung’s disease have been developed and modified, being associated with extensive surgical procedures involving laparotomy, resection of affected segment of colon and descensus of the functional bowel to the anus. Conventional surgery was followed by complications, although the incidence of early and late postoperative complications has significantly decreased. Transanal mucosectomy was practiced as a part of conventional and laparoscopic assisted pull through procedures. The latest promising achievement is entirely transanal pull-through procedure, described by De la Tore–Mondragon and Ortega–Salgado: entirely transanal endorectal pull–through, thus avoiding both laparotomy and laparoscopy. Due to minimal invasive surgery it offers excellent results. During a five year period (2003–2008), 24 patients were operated with this technique. The recovery was uneventful in all patients. This study adds valuable information of meticulous details of the new, still developing technique with the emphasis on controversies about surgical complications reported in multicentric studies.

Key words: Hirschprung’s disease, entirely transanal endorectal pull–through.

INTRODUCTION

A functional intestinal obstruction in Hirschprung’s disease (congenital megacolon) is caused by the absence of ganglion cells in the wall of colon. Its incidence is about 1: 5 000 live births. In more than 75% cases the recto sigmoid colon is affected. In spite of numerous surgical techniques and their modifications, the surgical treatment of Hirschprung’s disease remains controversial in terms of choosing optimal technique to avoid both early and late postoperative complications (incontinence, soiling, staining, strictures and enterocolitis), the incidence of complications being higher in multicentric studies.

Well known and most widely used classic techniques were introduced decades ago by Swenson, Duhamel and Soave, with subsequent numerous modifications of these three basic procedures. Laparoscopic surgery has been introduced lately in the treatment of Hirschprung’s disease, but nevertheless, although less invasive it is also based on classic surgical techniques. Transanal mucosectomy was practiced for many years as a part of conventional and laparoscopic assisted pull–through procedures for Hirschprung’s disease. The turning point in the surgical approach emerged in 1998 when De la Tore–Mondragon and Ortega–Salgado published the latest development – entirely transanal endorectal pull–through technique. Since then the technique has been widely accepted by pediatric surgeons. Few extensive multicentric studies have been published since then; most of the initial experiences with this technique were reported from single institutions with relatively small number of patients. Nevertheless, multicentric studies published so far reported uneven results due to differences in many parameters (age of patients, clinical manifestations, neglected and unrecognized disease, different preliminary procedures, i.e. protective colostomy and uneven surgical experience with conventional techniques). In multicentric studies higher incidence of intraoperative and postoperative complications were reported. They were mainly caused by the pitfalls of this sophisticated technique.

Therefore, any new reports from a single institution add important hints and peculiarities that might help in improving operative results and decrease unnecessary complications. De la Tore–Mondragon and Ortega–Salgado technique was implemented in our institution in 2003. During a five-year period (2003–2008) it was used in 24 patients. The authors of this paper have successfully demonstrated the technique in various major pediatric surgery hospitals in this country.
The purpose of this preliminary report was to describe in details the operative technique and to evaluate the pitfalls and the results of surgery based upon long follow up. Being the study from a single institution with uneventful full recovery of all patients, another aim was to evaluate possible causes of higher incidence of complications reported in multicentric studies.

MATERIAL AND METHODS

During a five-year period (2003 – 2008) 24 children were operated for Hirschprung’s disease with new entirely transanal approach technique. The following data were collected to elaborate the study:

1. Sex and age at diagnosis.
2. Previous procedures, if performed.
3. Anamnestic data, diagnostic procedures and preoperative regimen.
4. Detailed description of the procedure with emphasis on pitfalls that might compromise successful surgery, and operation time in minutes.
5. Early and late postoperative complications reported in multicentric studies (perianal excoriations, enterocolitis, anastomotic stricture, cuff abscess, rectal prolapse, anastomotic leak, constipation).
6. Outcome during long- term follow – up in terms of function (incontinence, soiling, staining, enterolitis and stricture of the anus).

RESULTS

1. Twenty four patients were operated with new De la Tore – Mondragon and Ortega – Salgado technique (21 male and 3 females – a well known ratio of more than 4 : 1 higher incidence in males, that has not been explained so far). Associated anomalies were detected in one patient (Down syndrome and congenital heart disease, i.e. atrioventricular canal successfully operated at the age of 4 months). Familial incidence was observed in one case; elder brother and sister in whom surgery was performed at the age of 6 and 3 months, respectfully. The mean age at the time of diagnosis and surgery was $5.3 + 3.1$ months. The youngest patient was operated in the neonatal period (25 days).

One male patient aged 12 years, with unrecognized and neglected illness, was excluded from statistical calculation of the mean age, being an exception in terms of manifestation of Hirschprung’s disease.

2. Five patients were referred to our institutions from elsewhere without definitive diagnosis of Hirschprung’s disease and therefore no preliminary procedures, i.e. colostomy were done.

3. Anamnestic data were typical in all children: delayed evacuation of meconial stool ($n=24$), abdominal distension ($n=24$), intestinal obstruction ($n=14$), failure to thrive ($n=5$) and severe enterocolitis in one case due to unrecognized Hirschprung’s disease. Diagnostic procedures included barium enema with definitive diagnosis being established with ex tempore biopsies of involved colon.

FIGURE 1.
A. RECTAL MUCOUS CYLINDER DISSECTED (ABOUT 6 CM). B. AGANGLIONIC COLON DISSECTED AND PULLED THROUGH SEROMUSCULAR CYLINDER (BLOOD VESSELS CAREFULLY LIGATED)

FIGURE 2.
DISSECTION OF RECTAL MUCOUS CYLINDER.

The patients were adequately prepared for surgery with normal saline enemas given twice a day. Newborns are particularly susceptible to the risk of iatrogenic colon perforation and enemas were given with utmost care by well-trained nursing staff.

Antibiotics were given 24 hours before surgery (amikacin, ampicilin and metronidazol) by intravenous route, and continued after the surgery for five to seven days.

4. The patient is placed in supine position with elevated pelvis. We routinely introduce urinary catheter into the bladder, although some authors think that this procedure is unnecessary. We are convinced that urinary catheter protects the urethra from accidental damage during the entire dissecting procedure, particularly by less-experienced surgeons.

Entire transanal one – stage endorectal pull – through surgical approach is demonstrated in Figure 1 A and 1 B. 
The rectal mucosa is incised circumferentially within the anus at the level of 0.5 to 1 cm above the dentate line. Using blunt dissection, the mucosa is dissected in the submucosal plane with constant but mild traction to facilitate the dissection of mucosal tube of 5 – 6 cm (Figures 1 A and 2). When transanal mucosectomy is completed, the seromuscular cuff is circumferentially incised to expose the sigmoid colon which is mobilized by ligating or cauterizing blood vessels.

The rectal mucosa is incised circumferentially within the anus at the level of 0.5 to 1 cm above the dentate line. Using blunt dissection, the mucosa is dissected in the submucosal plane with constant but mild traction to facilitate the dissection of mucosal tube of 5 – 6 cm (Figures 1 A and 2). When transanal mucosectomy is completed, the seromuscular cuff is circumferentially incised to expose the aganglionic colon, determined by multiple ex tempore biopsies (Figure 1B and 3). The length of dissected aganglionic colon varied from 10 to 35 cm (mean 20±7.3 cm) – in the exceptional case 54 cm were dissected in a 12-year-old patient (Figure 4).

The dissection of aganglionic colon is a meticulous procedure with a pitfall to damage blood vessels with their retraction upwards and severe bleeding impossible to control through narrow operative field i.e. through muscular cylinder. Such incidences demand an operative conversion to one of classic techniques involving laparotomy. Nevertheless, in experienced hands this severe surgical complication can be avoided, as well as other operative incidences – damage to vaginal wall and urethra9,10,11,12.

The Duration of the procedure in this series varied from 60 to 180 minutes (mean 90.4±17 minutes), the fact that speaks for itself that this is still not a routine operation even in experienced hands, fully accustomed to conventional techniques.

After the complete dissection of aganglionic bowel, uninvolved colon is pulled down the muscular cylinder and resected in horizontal plane (Figures 1B and 4). Circular anastomosis between the normally innervated colon and remaining mucosa is done above the dentate line, i.e. 0.5 – 1 cm above the mucocutaneous line. The level of circular rectoanal anastomosis is of utmost importance; if it is done distal to the dentate line it may damage subtle nerve endings causing functional complications (incontinence, soiling and staining). The space between dissected muscular cylinder and pulled-through normal bowel is drained. Another drain is inserted within the anus. Feeding was commenced after only 24 hours and drains removed after 2 – 4 days.

Routine anal dilatation began the eight days after surgery, this fact being very important to prevent both the stricture of circular anastomosis and the spasm of external sphincter. It also improves the function of descended bowel to resume its propulsive action.

5. The patients did not develop most commonly published complications, i.e. anal stenosis, fistula or abscess in the plane of dissection. Mild form – grade I and II of enterocolitis was observed in two patients (the first cases operated with the new technique).

Both patients recovered uneventfully after the application of enemas. Other insignificant postoperative events included mild diarrhea, slightly elevated body temperature, and mild distension of the abdomen. In the end, the recovery was uneventful in all patients. The average stay in the hospital was 10 days. At the time of discharge from the hospital the parents were instructed to recognize possible complications in operated children (constipation and enterocolitis).

6. The follow-up time was between 3 months and 5 years, mean 26 months8,6. Controls were done on monthly basis for the first three months, then every 3 months, and finally twice a year. All patients had normal defecation,
without enemas for constipation. There were no late post-operative complications (recurrent constipation, rectal prolapse, anal stricture and enterocolitis).

**DISCUSSION**

Entirely transanal approach for surgical treatment of Hirschsprung’s disease is relatively new procedure compared with many decades of surgical treatment based on various modifications of three basic techniques introduced by Swenson, Duhamel and Soave. All conventional techniques were a three – stage procedures: colostomy, with later resection of aganglionic colon and finally, the closure of colostomy.\(^1\)\(^2\)\(^3\)\(^4\)

Successful and uneventful surgery with decade’s long experience with classic techniques encouraged the surgeons to apply laparoscopic surgery without preliminary protective colostomy. Nevertheless, even less invasive laparoscopic approach was inevitably associated with entering abdominal cavity with intraoperative, early and late complications.\(^5\)\(^6\)

Entirely transanal approach is accepted as the method of choice by most pediatric surgeons due to its numerous advantages: low incidence of surgical incidents and complications in connection with laparotomy, intestinal obstruction due to adhesive bands, wound dehiscence and infection and intrabdominal abscesses.\(^1\)\(^2\)\(^3\)\(^4\)\(^5\) The risk to damage pelvic organs during colon dissection is minimal, as well as the damage of anal sphincter, particularly in experienced hands and meticulous dissection in well established and controlled dissection plane.

Dissection of mucosal tube is a demanding procedure in neonates and young infants because of possible disruption of mucosal cylinder. Limited and small operative field demands every precaution to avoid intraoperative incidents. The length of mobilized aganglionic colon to the transitional zone is most safely confirmed by ex–tempore biopsies, although some authors determine it on gross appearance at the level of transitional zone.\(^1\)\(^2\)\(^3\)\(^4\)\(^5\) We are strongly convinced that this approach is unsafe, unrespectable of surgical experience and obvious difference in the thickness of colon at transitional zone.

Transanal procedure is not advisable in children who had either preliminary colostomy or perforation of colon because of unavoidable adhesions which make the transanal dissection of colon unsafe or practically impossible. Endorectal technique can be applied in most of the patients with aganglionic rectosigmoid colon. Another advantage is the possibility to apply the technique in neonates.\(^1\)\(^2\)\(^3\)\(^4\)\(^5\) The limiting factor is the length of aganglionic colon because transanal dissection of aganglionic colon is determined by its anatomical mobility.

Dissection of longer proximal segments is augmented by laparoscopy.\(^5\) Another limiting factor may be marked hypertrophy and dilation of the colon proximal to transitional level in older patients. The problem is obvious – both dissection and pull through procedure are difficult having in mind limited diameter of seromuscular cuff. Therefore, the surgeons are advised to judge the optimal moment of conversion to laparotomy and continue the operation with conventional procedures.\(^3\)\(^4\) Stubborn persistence on transanal approach may cause serious intraoperative incidents, i.e. severe, uncontrollable bleeding. Therefore, an early diagnosis of Hirschprung’s disease in neonatal period is of the utmost importance to make entire transanal approach easier to perform.

Furthermore, early application of enemas both decreases the risk of complications and adequately prepares the patient for safe endorectal procedure.\(^9\)\(^10\)\(^12\)\(^14\) The timing of the surgery in infants on enema regimens depends on patient’s condition and the judgment of the surgeon. The younger the patient, the procedure is easier to perform, i.e. before marked dilatation of ganglionic colon proximal to the transitional zone is developed. Limited operative field and subtle anatomic structures in neonates and young infants are therefore not considered as limiting factors for endorectal pull – through procedure, providing that all patients are adequately prepared for the surgery.\(^5\)\(^10\)\(^12\)\(^14\)

**CONCLUSION**

Entire transanal pull – through for the surgical treatment of Hirschsprung’s disease was performed in 24 patients in our institution. Due to low natality rate and low incidence of the disease, the number of patients from a single institution is significant. There were no intraoperative incidents or complications. Good functional result is achieved in all patients i.e. regular defecation without laxatives or enemas. Uneventful outcome in all patients can be attributed to decades – long experience with conventional Soave modified technique or Duhamel technique, simultaneously performed in the same institution.

Large number of patients with Hirschprung’s disease operated with conventional techniques within the last 30 years exceeds 300 cases. We believe that this is the crucial fact for uneventful recovery and absence of reported early and late complications because an experienced surgeon with conventional techniques can perform and new technique fully aware of details and pitfalls that might compromise new surgical procedure. Therefore, although multicentric studies offer large number of patients, adding significant number of higher incidence of complications impossible to observe in small groups of patients, reports from single institutions are still highly valuable.

**SUMMARY**

**TRANSANALNA TEHNIKA ZA HIRURŠKO LEČENJE HIRSCHRUPNOVE BOLESTI – PRELIMINARNO SAOPŠTENJE**

Postoje brojne operativne tehnike i njihove modifikacije za lečenje Hirsprunrove bolesti. Većina hirurških tehnika zasniva se na ekstenzivnim hirurškim proce-
durama - laparotomija, resekcija zahvaćenog dela kolona i spuštanje funkcionalnog dela creva do anusa. Konven-
cionalne hirurške tehnike bile su često praćene komplikacijama, iako je incidencija ranih i kasnih postoper-
tivnih komplikacija znatno smanjena. Transanalna mukozektomija je primjenjivana kao deo konvencionalnih i la-
parskopskih pull – through procedura. Poslodaja metod
koja mnogo obećava je potpuni transanlni pull – through bez laparotomije ili laparoskopije. zbog minimalne invazivnosti ovom tehnikom postižu se odlični rezultati. tokom petogodišnjeg perioda (2003-2008) operisano je 24 bolesnika primenom ove hirurške tehnike. Oporavak posle operacije je kod svih bolesnika protekao bez komplikacija. ovim prethodnim saopštenjem obuhvaćene su korisne informacije o detaljima relativno nove hirurške tehnike koja se još uvek razvija i usavršava, sa naročitim osvrtom na kontroverze o hirurškim komplikacijama koje su objavljene u multicentričnim studijama.

 Ključne reči: hiršprungova bolest, potpuni transanlni endorektalni pull – through.

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