Laparoscopic-assisted Soave operation for the treatment of Hirschsprung disease in children: 5 years of experience

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Abstract

Purpose The purpose of this study was to summarize the clinical experience of the laparoscopic-assisted Soave operation for the treatment of Hirschsprung disease in children.

Methods In total, 186 children with Hirschsprung disease participated in this study from January 2014 to January 2019. The Soave operation was used to treat Hirschsprung disease with laparoscopic assistance. Symptoms and signs were followed up at one week, one month, three months, six months, one year and every 1-2 years after the first year.

Results All 186 children underwent laparoscopic surgery successfully, and none progressed to open surgery. During hospitalization and follow-up, there were 49 patients with complications, including 1 patient with an anastomotic leakage, 1 with an anal stricture, 5 with constipation recurrence, 5 with dirty defecation, 22 with enterocolitis, and 15 with perianal erosion. There were no complications such as abdominal bleeding, abdominal infection, ureter injury, adhesive intestinal obstruction, anastomotic stricture, incontinence.

Conclusion The laparoscopic-assisted Soave operation is a safe and feasible method for the treatment of Hirschsprung disease in children. This method has the advantages of less trauma and good cosmetic effects.

Introduction

Hirschsprung disease is one of the most common congenital digestive tract malformations in paediatric surgery, with an incidence of approximately 1/5000[1-3]. The main symptoms are delayed defecation, vomiting, progressive abdominal distension and constipation. If timely and effective treatment is not carried out, poor digestive system function can result, which affects growth and can even lead to death[4]. Surgical treatment is often needed in children with megacolon, and traditional surgical methods include the open operation of Duhamel, Swenson and Soave, with great trauma[5,6].

With the development and application of laparoscopic surgery in paediatric surgery and the continuous improvement of megacolon surgery, the operation for Hirschsprung disease has gradually evolved to laparoscopy-assisted combined with transanal anal pulling. As early as 1994[7] and 1995[8], Smith and Georgeson reported on the Duhamel and Soave procedures assisted by laparoscopy. Subsequently, the Soave operation with laparoscopic assistance has been widely used in the clinic, and remarkable results have been achieved[9-11]. We retrospectively analysed clinical data for 186 children with Hirschsprung disease who underwent the Soave operation assisted by laparoscopy in our hospital over 5 years to summarize the experience and clinical efficacy of this approach.

Methods

Patients
We retrospectively analysed the clinical data of 186 children with Hirschsprung disease in our hospital from January 2014 to January 2019, including preoperative, intraoperative, postoperative and follow-up data. Patients met the inclusion criteria if they underwent the Soave operation assisted by laparoscopy. Patients were excluded from this study if they 1) underwent other surgical methods 2) had severe liver and kidney dysfunction and complex congenital heart disease or 3) refused to sign the consent form for surgery or refused to comply with the follow-up schedule.

All the children had symptoms of delayed defecation and long-term or repeated constipation. Hirschsprung disease was clearly diagnosed by transanorectal manometry, barium colonography, intraoperative frozen pathology and postoperative paraffin pathology examination. Among the patients, 135 patients had the common type, 43 the long segment type, and 8 the total colon type. There were 122 males and 64 females. Age ranged from 2.3 months to 6.3 years, and the weight ranged from 3.5 kg to 26.6 kg (Table 1). A routine clinical examination was performed before the operation, including an electrocardiogram, chest radiography, cardiac colour Doppler ultrasound and blood examination.

**Technology**

**Preoperative preparation**

Normal saline was used to clean and wash the intestines for 7-10 days before the procedure.

**Surgical method**

After successful anaesthesia, each patient was placed in a flat position; routine surgical field disinfection was performed, and the lower extremities were aseptically clothed by package isolation. A pneumoperitoneum needle was injected into the abdominal cavity at the lower edge of the umbilical part, and an artificial pneumoperitoneum was established by slowly injecting CO₂ gas. The pneumoperitoneum pressure was generally maintained at 1.2-1.6 kPa. After the pneumoperitoneum needle was removed, a 5-mm inner-diameter laparoscope was placed at the puncture point, and operation forceps were placed at the intersection of the outer edge of the left rectus abdominis muscle and the umbilical horizontal line and the right lower abdomen. The extent of intestinal lesions was examined by laparoscopy to determine the spasmodic segment and the distal and proximal transitional segments. The serous muscle tissue of the upper rectum was cut and sent for intraoperative frozen biopsy, and the pathological report showed that the submucous and intermuscular ganglion cells were diagnosed as "congenital megacolon". The area from the lesion section and the distal end to the upper section of the rectum was retroperitoneal to 1-2 cm, the proximal end to the external appearance was soft, and the size of the intestinal tube was close to that of the normal intestinal tube. The proximal biopsies were taken from the proximal normal intestine. The sarco-muscular tissue of the segment was cut and sent for intraoperative frozen biopsy, and submucous and intermuscular ganglion cells were reported pathologically. After fully releasing the spleen curvature of the colon, the left side of the peritoneum, the lateral ligament of the descending colon, the lateral ligament of the sigmoid colon, and the proximal part of the examination could be stretched to the pelvic floor without tension. A length of 5-10 cm of "healthy"
colon with some ganglionic cells was resected. Then, we drained the gas from the abdominal cavity and began the anal surgery. From the dentate line of the rectum, the posterior wall was 0.8 cm, and the anterior wall was 1.5 cm; the rectal mucosa was cut in the oblique ring, and the rectal mucosa was free to the proximal end to the abdominal cavity. We circularly cut the muscle sheath and resected part of the rectum muscle sheath in the posterior wall with the V type. The intestines were pulled out from the abdominal cavity with no tension and no torsion. At the proximal end of the examination, the serous layer of the lower segment of the colon was sutured with the stump of the rectal muscle sheath using an absorbable 4-0 line. The colon was cut off at the 0.5-cm distal end of the anastomosis, and intermittent suturing was performed between the severed end of the colon and the rectal mucosa. The artificial pneumoperitoneum was re-established; no active bleeding was found in the abdominal cavity, and the pulled-out colon was not reversed. The pneumoperitoneum gas was discharged, and the abdominal incision was closed.

Postoperative management

After recovery of intestinal function, anal exhaust and defecation, the children began to take fluids and gradually returned to a normal diet after 5 days. Anal dilatation began at 14 days after the operation, once a day, and was fixed for 15 min to 20 min, increasing by 1 calibre (1 mm) every week and lasting for 3 months.

Postoperative follow-up

The children were followed up by telephone and outpatient service. The follow-up times were at one week, one month, three months, six months, and one year after the operation, and then every 1-2 years thereafter. Instances of constipation, faecal incontinence, defaecation, enteritis, etc., were recorded.

Results

All 186 children underwent laparoscopic surgery successfully, and no cases progressed to open surgery. During hospitalization and follow-up, there were 49 patients with complications, including 1 with anastomotic leakage, 1 with anal stricture, 5 with constipation recurrence, 5 with dirty defecation, 22 with enterocolitis, and 15 with perianal erosion. The patient with the anal stricture, which was caused by the lack of time for the parents to dilate the anus, was cured by correct and reasonable anal dilatation after 1 year. For the 5 patients with constipation recurrence, 2 were cured by conservative treatment, and 3 were cured by repeat open operation for the transition zone. The defecation symptoms of the 5 children with dirty defecation gradually disappeared with age and long-term anal sphincter exercise training. Children with enterocolitis were cured after conservative treatment, such as anti-infective use, clean enaemas, and probiotics. The children with perianal erosion were cured after strengthening perianal nursing, keeping the perianal skin dry and clean, protecting the perianal skin with external drugs, taking oral intestinal convergent drugs and reducing stool moisture.
Discussion

Resection is the main treatment for Hirschsprung disease, the purpose of which is to remove the diseased intestinal canal and pull the intestinal canal innervated by the normal nerve to the anus for anastomosis to maintain normal function of the anal sphincter and achieve the purpose of continuity of digestive tract reconstruction[12]. In recent years, the one-stage radical Soave operation of the transanal megacolon[13] has been widely carried out because laparotomy is avoided; there is also less trauma, less bleeding and rapid postoperative recovery. However, it is only suitable for the short segment type and some infants with the common type of megacolon. The application of laparoscopy can resolve the technical limitations of the Soave transanal megacolon and reduce the trauma of laparotomy, which highlights its minimally invasive features.

Although laparoscopic surgery has many advantages, because of the small abdominal cavity in children, abdominal distension often affects the laparoscopic visual field, resulting in abdominal organ injury, defective intestinal tube judgement, and normal laparoscopic operation[14]. We took the following measures to reduce abdominal distension and the difficulty of the operation, which ensured a smooth operation and avoided or reduced the conversion to laparotomy. First, we chose an experienced anaesthesiologist to avoid prolonged mask oxygen supply and repeated tracheal intubation. If there is obvious gas accumulation in the gastric vesicle, we can properly adjust the position of the gastric tube, keep the gastric tube unobstructed, and expel the gas from the stomach. Second, insertion into the anal canal or adult gastric canal through the anus and insertion of the narrow segment into the dilated segment was performed to discharge the intestinal gas. Third, the small intestine often accumulates gas dilatation in the total colon type megacolon. The epidural catheter can be inserted through the abdominal wall to the dilated small intestine to eliminate the gas in the dilated small intestine and eliminate abdominal distension. Through the above measures to eliminate abdominal distension, laparoscopic surgery was successfully completed for all the children in this study, and no cases were converted to open surgery.

Enterocolitis is the most common and serious postoperative complication of Hirschsprung disease, with an incidence of 2-33%[15,16]. Some scholars believe that the occurrence of enterocolitis is related to incomplete colorectal obstruction[17], but we have observed that enterocolitis still occurs despite a smooth operation for most children, standard anal dilatation after the operation, and no obvious stricture or obstruction at the distal end of the colon. Therefore, we believe that in addition to colon obstruction, it is important to have low immunity in the body or intestine, to reduce surgical trauma and to avoid an imbalance of the intestinal flora. All the children in this study were cured after conservative treatment, and some of the children with recurrent enterocolitis recovered gradually with age and improvement in immune function.

Dirty defecation is a common postoperative complication of Hirschsprung disease. The main reasons are injury to the anal sphincter or excessive traction of the anus during the operation. When the colon is pulled out of the anus, the anal sphincter is damaged, leading to dysfunction of the anal sphincter. The
occurrence of some dirty defecation is also related to congenital anal sphincter dysplasia\[18\]. Therefore, the activity in the perineum should be gentle during the operation to avoid excessive traction of the anus; injury of the levator anal muscle should also be avoided during laparoscopic free retroflexion of the intestine. In this study, the children with dirty defecation were not found to have congenital dysplasia or congenital loss of the anal sphincter by MRI, and all were cured after exercise training of the anal sphincter.

The causes of recurrence of constipation after the operation are as follows\[19,20\]. 1. The rectum muscle sheath without ganglia was retained too long. There was no incision or an insufficient incision of the posterior wall of the rectal muscle sheath during the operation. 2. The resection of the diseased intestine was insufficient, from a long segment megacolon to a short segment or ultra-short segment megacolon. 3. Secondary ganglion cells develop poorly due to improper operation, proximal intestinal injury, or ischaemia. 4. During the operation, the abdominal cavity was widely separated, and the blood vessels were damaged, resulting in spasm caused by insufficient blood supply to the internal sphincter and anal stricture, resulting in constipation recurrence. 5. Enterocolitis is also an important cause of constipation recurrence. In this study, 5 patients with constipation recurrence and 2 with conservative treatment improved after the operation. One patient had recurrent, persistent constipation after the surgery, and radiography showed colon dilatation. Considering that the resection scope was not sufficient, we resected the dilated segment. In one case, frozen pathology revealed a long segment megacolon; the whole colon was pathologically confirmed in paraffin-embedded samples after the operation, and the radical extubation operation was performed again. One case of Hirschsprung disease was complicated with megacolon-like disease with insufficient intestinal resection, resulting in a recurrence of constipation, which was cured by reoperation. Therefore, the cause of abnormal defecation after the operation must be found. After excluding anastomotic stricture, it should be clear whether development of the intestinal nerve is normal and whether it is complicated by megacolon. It has also been suggested that the pathological diagnostic criteria and pathologists’ experience should be emphasized in the radical resection of megacolon.

There are several limitations of our study. First, this was a single-centre study, and more research from multiple centres is needed to assess the effectiveness and complications of this technique. Second, this study was a retrospective review without a control group.

**Conclusion**

In conclusion, laparoscopic-assisted Soave surgery is a safe and feasible method for the treatment of Hirschsprung disease in children. The cosmetic results are impressive, and the follow-up results are promising.

**Declarations**
Ethics approval and consent to participate This study was approved by the ethics committee of Fujian Maternity and Child Health Hospital and strictly adhered to the tenets of the Declaration of Helsinki. In addition, all patients’ guardians signed an informed consent form before the operation.

Consent for publication Not Applicable

Availability of data and materials The datasets used and analysed during the current study are available from the corresponding author on reasonable request.

Competing interests The authors declare that they have no competing interests.

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Authors’ contributions LMK and FYF designed the study, collected the clinical data, performed the statistical analysis, participated in the operation, and drafted the manuscript. ZB, LY, LOM, BJX and WDM participated in the operation and revised the article. All authors read and approved the final manuscript.

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**Table**

Table 1. Clinical data of the patients in this study
| Item                                                   |       |
|--------------------------------------------------------|-------|
| Number of patients                                     | 186   |
| Boys/Girls                                             | 122/64|
| Age                                                    | 7.6month(2.3months-6.3years) |
| Weight                                                 | 6.9kg-3.5kg-26.6kg       |
| Common type/Long segment type/Total colon type         | 135/43/8 |
| The operative time                                     | 3.1(2.3-4.6)h             |
| The amount of bleeding                                 | 5(2-12)ml                   |
| The postoperative time to eating                       | 50(30-76)h                  |
| The postoperative hospital stay time                   | 7(5-10)d                    |
| Scar length                                            | 0.8(0.6-1.2)cm             |
| Other anomalies                                        |       |
| Simple congenital heart disease                        | 22    |
| Renal hydronephrosis                                  | 6     |
| Hypospadia                                            | 2     |
| The follow-up time                                    | 3.5years(3months-5years)   |