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Soave transanal one-stage endorectal pull-through in the treatment of Hirschsprung's disease of the child above two-year-old: A report of 20 cases

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ABSTRACT

Background: The definitive treatment of Hirschsprung's disease is the removal of the aganglionic bowel by a pull-through surgery. In most cases, this surgery is performed in infancy or in the neonatal period as presentation in older children and adulthood is uncommon. **Materials and Methods:** It is a retrospective study of 20 patients above two-year-old who underwent a transanal Soave one-stage endorectal pull-through procedure for Hirschsprung's disease between January 2002 and December 2010. **Results:** Twenty patients were recruited in this study. Fourteen were males and six were females. Patient ages ranged from 2 to 14 years (median age: five years and three months). All patients presented with persistent constipation and abdominal distension. Two of them had an intestinal obstruction that required colostomy. Ten patients (50%) had a recto-sigmoid Hirschsprung's disease. All patients were operated on using a Soave one-stage endorectal pull-through procedure. The laparoscopy was necessary during the pull-through in three cases. The average duration of the intervention was 240 minutes. That represents almost the double of the duration of the same procedure in newborns and infants in our department (130 minutes). Early postoperative complications included one case of anastomosis leakage and one case of intussusception. Late postoperative complications were perineum irritation in five cases (25%), anal stenosis in four cases (20%) and enterocolitis in one case (5%). None of our patients developed fecal incontinence. Soiling was reported in four cases (20%). There was no death. **Conclusion:** Soave transanal one-stage endorectal pull-through is safely feasible in children of more than two years of age. Laparoscopy may be necessary whenever there are difficulties in the pull-through.

Key words: Complications, difficulties, Hirschsprung disease, infant, laparoscopy, soave procedure, surgery

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INTRODUCTION

The diagnosis of Hirschsprung disease is almost done in the neonatal period. If not done, older infants and children typically present with chronic constipation and abdominal distension.

The basic principle for the definitive surgical therapy is resection of the aganglionic bowel followed by a pull-through of ganglionic bowel down to the anus. Soave transanal one-stage endorectal pull-through is at present the most commonly used technique in neonates and infants.

In children, most surgeons have some difficulties in this procedure, also there are few reports in literature about this technique. This study aims to improve knowledge about this technique which may lead to further refinements of the surgical techniques and better treatment of these children.

MATERIALS AND METHODS

From January 2002 to December 2010, 20 children aged above two years old and diagnosed with Hirschsprung's disease underwent a Soave one-stage endorectal pull-through procedure in the department of pediatric surgery of Monastir/Tunisia.

Patients that were operated before the age of 2-year-old, and then required a redo endorectal pull-through, were included in this study.

The medical records of all patients were retrospectively reviewed. The collected data included demographic features, clinical presentation, age at diagnosis, preoperative investigations, level of aganglionosis, the particularities and technical difficulties during each intervention, pathology reports, complications and functional outcome (evaluated according to the score of Holschneider).[1]

In the same period of study, 90 patients under two years were operated by using the same technique.

The diagnosis of the Hirschsprung's disease was based on the barium enema showing a caliber disparity and preoperative rectal biopsy. Ano-rectal manometry was necessary in six cases where the contrast enemas were not clearly demonstrative.

All patients had a preoperative bowel preparation including digital rectal stimulations and rectal irrigations with saline solution 20 cc/kg two times per day. A preoperative antibiotic prophylaxis with amoxicillin and clavulanic acid was given in all cases. In a lithotomy position the abdomino-perineal area is prepared and draped. Stay sutures were placed around the ano-cutaneous junction to permit an eversion of the entrance of the anus. The mucosa was incised circumferentially just 1 cm (average) above the dentate line and a submucosal dissection was carried proximally. When the dissection extended to a point above the peritoneal reflection, the rectal muscle was incised circumferentially and the dissection carried along the outer wall of the rectal muscle. When the transition zone was reached, an intraoperative frozen section was performed. The colon is transected at the area that has been proved to have normal ganglion cells by the pathology examination. Coloanal anastomosis is completed using absorbable sutures.

In the cases where the aganglionic area extended beyond the sigmoid colon, an intraperitoneal laparoscopic dissection was performed.

RESULTS

A total of 20 children were included in this study: 14 boys (70 %) and 6 girls (30 %). The sex ratio was 2:3.

All patients presented with long-term constipation, except two cases which had a digestive diversion by colostomy before the Soave pull-through procedure.

A barium enema was performed in all cases. It showed the typical picture of Hirschsprung disease in 17 cases (85%) and demonstrated transition zone in the rectosigmoid region in half cases. For the remaining cases three cases (15%), barium enema showed no transition zone, but there was a marked rectocolic dilatation which was consistent with juxta-anal forms.

Ano-rectal manometry was performed in six cases 30% (three cases with juxta-anal forms and three cases with rectal forms on the contrast enema). The rectoanal reflex was absent in all six cases.

Anatomopathological examination confirms the diagnosis in all cases by demonstrating the absence of ganglion cells in the diseased segment.

A temporary *colostomy* was performed before the pull-through procedure in two cases. It was indicated because of a digestive perforation in one case, and a failure of nursing with intestinal obstruction in the other case.

The age at the time of the definitive pull-through surgery varied from two to 14 years, with an average of five years and three months.

No patient required a conversion to laparotomy. A laparoscopy was necessary in three cases (15%). It was decided:

- Before the pull-through to achieve the intra-abdominal and pelvic dissection in two cases with an aganglionosis that extended beyond the sigmoid colon.
- And during the pull-through surgery because of the retraction of the colon into the abdomen in one patient that has had a colostomy during six years. The laparoscopy was then used to help the mobilization of the intraperitoneal part of the colon. Extensive interloop colonic adhesions were found.

The average duration of surgery varied from 120 to 360 minutes, with an average of 240 minutes.

Intraoperative biopsies of the circumference of the bowel to be used for pull-through were performed in all cases, with histological confirmation of the presence of both ganglion cells and normal caliber nerves.

The length of the resected colon ranged from 7 to 32 cm (median, 17 cm). This measurement included

the aganglionic segment in addition to 3-10 cm of the dilated zone.

The length of agangionosis, based on the result of definitive anatomopathological examination, ranged from 5 to 29 cm (median, 11 cm).

The postoperative hospital stay ranged from 3 to 10 days (median, four days).

A digital rectal examination was performed 15-21 days after surgery to detect anastomotic stenosis and dilate it.

Early postoperative complications included:

- One case of an ileo-ileal intussusceptions which appeared three days after the surgery, it was reduced laparoscopically with good results.
- And a case of anastomotic leak probably due to an incongruence of the anastomosis, which required a surgical repair [Figure 1].

The late complications were dominated by:

- Irritation of the perineum in five cases (25%), with improvement after two to three months of local care (Vit A and D pomade).
- Anal stenosis in four cases (20%) that evolved favourably after digital or instrumental anal dilation.
- Postoperative enterocolitis was met in one case (5%). It was treated by antibiotics and enemas. The evolution was good and there was no recurrence.

To evaluate the functional outcome of our patients, we used the score of Holschneider.[1] No case of anal incontinence was reported.

The frequency of soiling was 20% (four cases). This can be explained by the dilatation of the colon above the aganglionic area. Soiling was improved by dietary advice, cognitive and behavioural therapy positively, and ano-rectal biofeedback using instrument-assisted exercises.

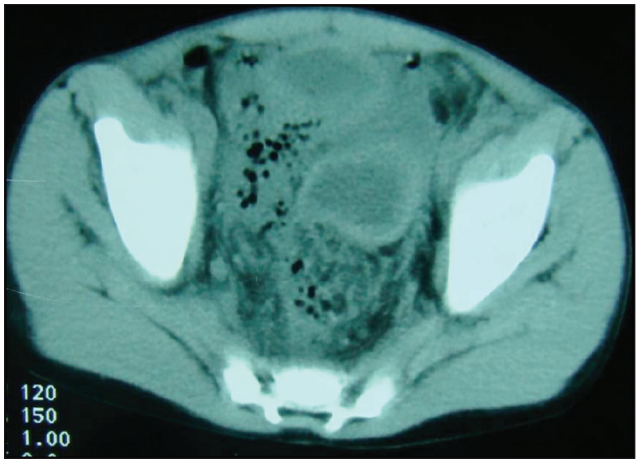


Figure 1: Abdominal tomodensitometry showing a pelvic abscess due to an anastomotic leak probably due to an incongruence of the anastomosis, which required a surgical repair

Postoperative constipation developed in two cases (10%). The first case was improved by medications and dietary measures. In the second case, the rectal digital examination showed an anastomotic stricture. The re-reading of the pathology specimens confirmed the presence of ganglionic cells in the surgical limit of the resection. We retained the diagnosis of a hypertony of the internal sphincter and the child had an internal sphincteromyotomy. The evolution was favourable.

DISCUSSION

Soave transanal one-stage endorectal pull-through is the most used technique in newborns and neonates for the treatment of the disease of Hirschsprung's disease thanks to its multiple advantages: bloodless easy mucosectomy, absence of any abdominal incision, easier control of the blood supply of the colon, less risk of damage to pelvic structures, a lower incidence of intraperitoneal bleeding and adhesion formation, minimal pain, early discharge, lower total cost, satisfactory cosmesis and lower rate of complications.[2-7]

In the literature there are few reports concerning this technique in patients aged above two years old.

Based on our experience, Soave transanal one-stage endorectal pull-through is feasible in children aged above two-year-old and it has some particularities that need to be taken into account:

- The chronic accumulation of stools causes the inflammation, the ulceration of the mucosa and repeated episodes of subclinical enterocolitis which are responsible for the development of fibrous adhesions between the mucosa and the submucosa [Figure 2].
- The mesenteric vessels in this age group are larger and may bleed in spite of electrocoagulation[6,8] [Figure 3].
- The difficulty of displaying the very dilated colon, in spite of the opening of the muscular cylinder [Figure 4].
- Anastomotic incongruence that may require the resection of the dilated proximal colon in order to have a near normal calibre colon also several stitches may be required.[8-10]

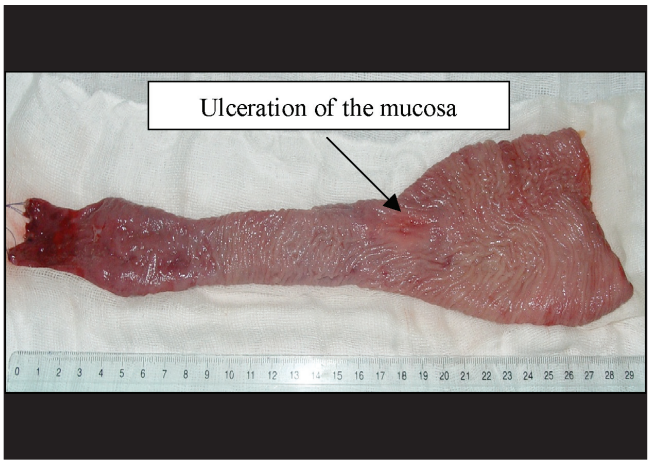


Figure 2: Ulceration of the mucosa

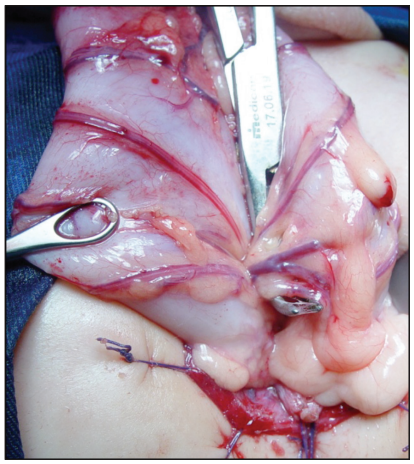


Figure 3: Large vessels in the old child with a higher risk of bleeding

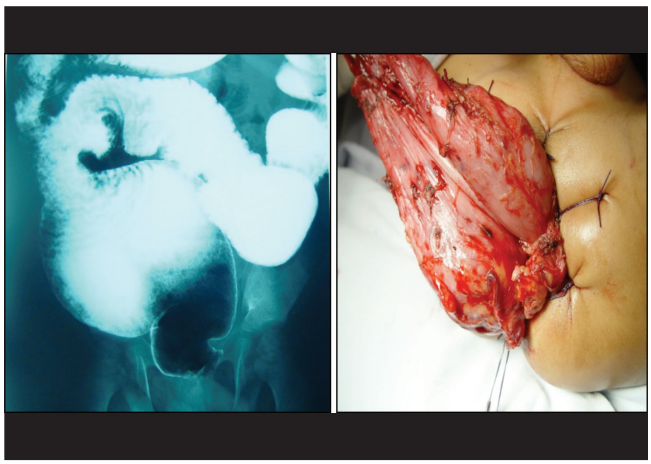


Figure 4: Very dilated colon which can causes difficulties in its mobilisation

The Soave technique requires:

- A preoperative bowel preparation by a series of enemas to empty the bowel of stool and reduce the dilatation of the bowel.
- An operating time that is two times longer than in newborns and infants.
- A perfect colo-anal anastomosis to prevent the problems of incongruence and anastomotic leaks.

1. The most serious early operative complication is anastomosis leakage.[11,12]

   It can result from:

   - A tension on the anastomosis.
   - Inadequate preoperative bowel preparation.
   - A technical problem concerning the placement of sutures or an inadequate matching of luminal diameters.
   - Inadequate blood supply up to and including edge of the anastomosis.

   In this series, this complication was met in one case. It was rapidly diagnosed and demonstrated by abdominal CT scan. The patient was reoperated on: as the diagnosis was done very soon, we preferred a redo soave than to divert faeces by creating a stoma and reanastomosis later.

   One patient had intussusceptions during the immediate postoperative period. This complication was not reported in literature before.

   The main long-term complications were:

2. Perineum irritation that it is less frequent than in newborns and infants whose skin is particularly sensitive.[13]

   - Enterocolitis is more frequent in children under two-year-old due to an immaturity of the enteric nervous system.[14]

   On the functional level:

   - No patient presented anal incontinence in this series. According to the literature, this frequency varies from 1 to 10 %.
   - Soiling was estimated at 20% at in children above two years of age and 13% in neonates and infants. This can be explained by the dilatation of the colon above the aganglionic area. The review of the literature found that the frequency of soiling varies from 6 to 12%. It is managed with dietary measures, cognitive and behavioural therapy, and ano-rectal biofeedback. The long-term outcome of this treatment is favourable in most cases.

3. The postoperative constipation was seen in two of our patients (10%). In the first case, It was medically handled. In the second case, we retained the diagnosis of a hypertony of the internal and the child had an internal sphincteromyotomy. This complication was reported, in the literature in 9.5%.[11,12,15]

   - of the children with Hirschsprung disease who underwent Soave transanal one-stage endorectal pull-through regardless of the age group.

CONCLUSIONS

In children above two years of age with Hirschsprung's disease, Soave transanal one-stage endorectal pull-through was found to be safe applicable, and with low associated morbidity and mortality. We emphasize, in this age group, on the importance of preoperative preparation of the bowel and the experience of the surgeon. Laparoscopy should be associated whenever there are difficulties in the pull-through.

REFERENCES

1. Holschneider AM, Ure BM, Pfommer W. Innervation patterns of the rectal pouch and fistula In ano-rectal malformations: A preliminary report. J Pediatr Surg 1996;37:357-62.
2. Langer JC, Minkes RK, Mazziotti MV, Skinner MA, Winthrop AL. Transanal one-stage Soave procedure for infants with Hirschsprung disease. J Pediatr Surg 1999;34:148-52.
3. De La Torre L, Langer JC. Transanal endorectal pull-through for Hirschsprung disease: Technique, controversies, pearls, pitfalls, and an organized approach to the management of postoperative obstructive symptoms. Semin Pediatr Surg 2010;19:96-106.
4. Kim AC, Langer JC, Pastor AC, Zhang L, Sloots CE, Hamilton NA, *et al*. Endorectal pull-through for Hirschsprung's disease-a multicenter, long-term comparison of results: Transanal vs transabdominal approach. J Pediatr Surg 2010;45:1213-20.
5. Pratap A, Shakya VC, Biswas BK, Sinha A, Tiwari A, Agrawal CS, *et al*. Single-stage transanal endorectal pull-through for Hirschsprung's disease: Perspective from a developing country. J Pediatr Surg 2007;42:532-5.
6. Hadidi A. Transanal Endorectal Pull-Through for Hirschsprung's Disease: Experience with 68 Patients. J Pediatr Surg 2003;38:1337-40.
7. Vijaykumar, Chattopadhyay A, Patra R, Murulaiah M. Soave Procedure for Infants with Hirschsprung's Disease. Indian J Pediatr 2002;69:571-2.
8. Liu DC, Rodriguez J, Hill CB, Loe WA Jr. Transanal Mucosectomy in the Treatment of Hirschsprung's Disease. J Pediatr Surg 2000;35:235-8.
9. Langer JC, Seifert M, Minkes RK. One-stage Soave pull-through for Hirschsprung disease: A comparison of the transanal vs open approaches. J Pediatr Surg 2000;35:820-2.
10. Pratap A, Gupta DK, Shakya VC, Adhikary S, Tiwari A, Shrestha P, *et al*. Analysis of problems, complications, avoidance and management with transanal pull-through for Hirschsprung's disease. J Pediatr Surg 2007;42:1869-76.
11. Marty TL, Seo T, Matlak ME, Sullivan JJ, Black RE, Johnson DG. Gastrointestinal function after surgical correction of Hirschsprung's disease: Long-term follow-up in 135 patients. J Pediatr Surg 1995;30:655-8.
12. Marquez TT, Acton RD, Hess DJ, Duval S, Saltzman DA. Comprehensive review of procedure for total colonic aganglionosis. J Pediatr Surg 2009;44:257-65.
13. Wester T, Rintala RJ. Early outcome of transanal endorectal pull-through with a short muscle cuff during the neonatal period. J Pediatr Surg 2004;39:157-60.
14. Marty TL, Matlak ME, Hendrickson M, Black RE, Johnson DG. Unexpected death from enterocolitis after surgery for Hirschsprung's disease. Pediatrics 1995;96:118-21.
15. Little DC, Snyder CL. Early and late complications following operative repair of Hirschsprung's Disease. New York: Springer; 2008. p. 375-85.

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