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**Endoscopic management of sigmoid volvulus in children**

Parolini F *et al*. Sigmoid volvulus in children

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**Abstract**

Sigmoid volvulus (SV) is extremely uncommon in children and is usually associated with a long-standing history of constipation or pseudo-obstruction. An early diagnosis and management are crucial in order to prevent the appearance of hemorrhagic infarction of the twisted loop, avoiding further complications such as necrosis, perforation and sepsis. In patients with no evidence of peritonitis or ischemic bowel, treatment starts with resuscitation and detorsion of the SV, accomplished by means of sigmoidoscopy and concomitant rectal tube placement. The bowel is then prepared and surgery is undertaken electively during the same hospitalization. We report a detailed review of the literature focusing on technical details, risks and benefits of endoscopic management of SV in childhood.

**Key words:** Sigmoid volvulus; Contrast enema; Endoscopy; Children; Surgery

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**Core tip:** Authors provide a detailed review of the literature focusing on technical details, risks and benefits of endoscopic management of sigmoid volvulus in children.

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**INTRODUCTION**

Sigmoid volvulus (SV) is extremely uncommon in children and is usually associated with a long-standing history of constipation or pseudo-obstruction[1,2]. Patients with SV present redundant sigmoid colon with a narrow mesenteric attachment to posterior abdominal wall, allowing the close approximation of two limbs of sigmoid colon and making it prone to torsion around the mesenteric axis. Less frequently, predisposing factors are Hirschsprung’s disease and roundworm infestation, especially in smaller children. Presentations can range from acute to recurrent abdominal pain, often relieved by passage of stool or flatus[2,3]. An early diagnosis and management are crucial in order to prevent the appearance of hemorrhagic infarction of the twisted loop, avoiding further complications such as necrosis, perforation and sepsis[1-4]. If no signs of bowel ischemia and perforation are present, endoscopic decompression and detorsion of the volvulus has been proposed as the first step of treatment, followed by elective surgery with sigmoid resection and primary anastomosis[2,4]. We report a detailed review of the literature focusing on technical details, risks and benefits of endoscopic management of SV in children.

**REVIEW**

Multicenter studies on endoscopic management of SV in children are lacking. The initial PubMed search yielded 39 potentially relevant articles on the topic. Inclusion criteria were articles that reported original data on endoscopic management of SV in children younger than 18 years and they clearly reported the method of endoscopic treatment. Titles and abstracts of the identified publications were checked and reviewed against the predefined inclusion criteria, and afterward, the full text articles was reviewed[5]. Finally, 6 eligible articles were enclosed in the review, encompassing a total of 81 cases (Table 1)[1,2,6-9]. All but one studies were single institution case reports or case presentation (classes of evidence III and rating scales of evidence E)[5]. Only one multicenter study was found, encompassing 13 cases[1]. Another study provided a detailed retrospective review of 63 children with SV published in literature from 1940 to 1999[8]. Hirschsprung’s disease was associated in 13 out of 81 patients (16%). All patients of this series underwent endoscopic detorsion; the procedure was successful in 56 of cases (69%). Although this limited pediatric experience, evidence suggests that endoscopic management of SV should be considered the first step of treatment of these patients, followed by definitive elective surgery. Operative and technical details of endoscopic management thereby originated from a larger adult experience, as more than 1000 cases of endoscopic detorsion are reported[3,4].

**ENDOSCOPIC MANAGEMENT OF SV IN CHILDREN**

***Which patients should be endoscopically managed?***

All selected studies agree that emergency surgery is obviously indicated when the patient has clinical or radiological evidence or suspicion of peritonitis or perforation, which may clinically manifest as melanotic stool during anamnesis or rectal examination, guarding or rebound tenderness during abdominal examination[10]. In this subset of patients, the surgical procedure is chosen on the basis of the history, clinical presentation and intraoperative findings[10-12]. On the contrary, when signs of bowel ischemia and perforation are ruled out and a pediatric endoscopy service with high expertise or endoscipic guard with experience in pediatric procedures are available, endoscopic decompression and detorsion should represent the initial step of treatment in order to relief symptoms and to prepare the patient to semi-elective surgical exploration[1-6]. Surgery in emergency situations, when the general condition of the patient is suboptimal and the bowel is not prepared, is reported to carry higher complication rate[13].

***How should the patient be prepared?***

All selected studies agree that patients should actively be resuscitated by means of nasogastric suction and correction of fluid-electrolyte imbalance. Nasogastric intubation is necessary in order to allow gastric decompression, relief of symptoms and bowel rest and identification of the stomach on X-ray[1,10,11]. Broad spectrum antibiotic covering anaerobic bacteria should be administered immediately after admission. In patients with no evidence of peritonitis or ischemic bowel, water-soluble contrast enema is advisable before the endoscopy, in order to confirm the diagnosis and to rule out other causes of obstruction, such as intussusception[8]. Successful temporary reduction of SV by contrast enema is reported in up to 77% of the cases; moreover, enema could also facilitate preparation of both patient and bowel for subsequent endoscopy and surgery[8,9].

***What is necessary to perform a safe procedure?***

The procedure should be performed under general anesthesia in operating room[1]. This fact is different compared with adults, in whom the procedure could be safely performed under sedation in endoscopic suite. Different types of pediatric flexible colonoscopes less than 12-mm are commercially available. They are equipped with 3.2-mm biopsy channel, which allows the use of operative devices as biopsy forceps, snares and needles. Unfortunately, these instruments are more suitable for children 2 years and older (weight over 12-15 kg), and, as colonoscopes specifically designed for infants and toddlers do not exist, pediatric upper GI videoendoscopes can be used. It is assumed that is more difficult to study the sigmoid colon with these instruments, but their smaller diameter prevents excessive stretching of the intestinal wall, especially in newborns and infant. Nevertheless, in this series endoscopic management of SV was not attempted in children younger than 6 years old. Rigid colonoscopes should not be used, as in all but one selected studies[3] they are generally associated to higher risks of perforation and lower volvulus reduction successful rate[12-14].

***Endoscopic procedure***

Sigmoidoscopy is best performed with patient in the Sims or left lateral decubitus position. Hips and knees are partially flexed and the right knee is positioned above the left one[12-16]. The pediatric endoscopist should stand between the light source and the back of the patient. Digital rectal examination is advisable, in order to lubricate the anal canal, relax the rectal sphincter and give an initial assessment of the effectiveness of the bowel preparation. The lubricated tip of the scope should be gently introduced into the rectum by flexion of the right index finger, guiding it into the anus at a 90° angle. The less amount of air is insufflated, in order to avoid the stretching of bowel loops and to reduce the patient's discomfort after the procedure[16]. The evaluation of the colorectal mucosa should be performed during the withdrawal of the instrument. Liquid in the rectum should be aspirated *via* the sigmoidoscope for a clearer view. The sigmoidoscope is then advanced into the rectum under direct vision. The rectum is gently insufflated to provide good visibility and to facilitate identification of rectosigmoid junction, which represents the area of most difficulty during the examination. To overcome this step the endoscope should be advanced beyond the valve of Houston, then the tip should be deflect upwards and, with gentle clockwise torquing, slowly advanced beyond the rectosigmoid junction. Spirally twisted or converging colon mucosa (“whirl sign”) at the rectosigmoid junction indicates the distal point of torsional obstruction[10-12].The endoscope should be gently advanced through the apex of the converging mucosa into the dilated sigmoid colon. Ischemic changes of the mucosa or gangrene should be noticed and represent an absolute indication to discontinue the endoscopy and to convert to surgery[2-6].On the contrary, the management of children in whom endoscopic examination shows borderline ischemia is controversial[10-12]. Once the dilated sigma is decompressed and the endoscope is in the descending colon, endoscopic detorsion of the decompressed volvulus is obtained performing by clockwise rotation and shortening of the endoscope by the right hand. Only occasionally, the pressure of the air causes detorsion with reduction of the volvulus. If detorsion does not occur, the spiraling rectal mucosa is followed upward to the apex, and a soft rectal tube is passed up through this under direct vision[15,16]. The tip of the endoscope can also be used to apply a constant pressure at the apex of the twist, which can lead to detorsion and decompression[2]. A successful deflation is accompanied by a large amount of release of gas and liquid stool from the anus[1]. Eventually, rectal suction biopsies should be obtained, as Hirschsprung’s disease (HD) has been reported in up to 17% of cases of SV in infancy[8,17].

***Is rectal tube placement necessary?***

Evidence suggests that the placement of a rectal tube for 24-72 h helps to stabilize the patient further and prevents an early relapse of volvulus[1]. After the placement of a guide wire (0.035 inch), a multiple sideports guiding catheter is advanced through the endoscopic channel into the descending or transverse colon. Several devices are available and used in the adult setting for treatment of acute non-toxic megacolon, pseudo-obstruction and colonic strictures, including the 14 Fr Colon Decompression Set (Cook Inc, Bloomington, Indiana, United States) and 7 Fr, 8.5 Fr and 10 Fr Marcon Colon Decompression Set (Cook Inc). Endoscopic exchange was performed by gently pulling back the endoscope over the guidewire while advancing the guide wire. The drainage catheter was then advanced over the guide wire overcoming the point of the obstruction, and eventually the guidewire is removed through the drainage catheter[1,16,17]. Placement of a larger red rubber catheter per rectum alongside the scope is suuggested when colonic decompression kit is not available. When the tip of the catheter is visualized, biopsy forceps passed through the work channel of the scope are used to grasp the tip of the catheter and advanced it as far as necessary. The drainage catheter is taped over the perianal skin and should left in place for 1-3 d before surgery[1].

***The role of percutaneous endoscopic sigmoidopexy***

Described in the first time by Choi *et al*[18] in 1998, percutaneous endoscopic sigmoidopexy (PES) has been proposed in order to prevent recurrence of volvulus for elder patients who otherwise had contraindication for elective surgery and general anesthesia. PES is performed using the percutaneous endoscopic gastrostomy (PEG) technique. Nevertheless, as only one fixation point may be insufficient for preventing SV, Ito and colleagues reported PES with multiple fixation points in a 86-year-old patient with recurrent SV[19]. The sigmoid colon was fixed at six points to the abdominal wall using non-absorbable sutures, with the fixation knots buried subcutaneously, obviating the need for suture removal. Pinedo et al reported two patients in whom sigmoidopexy was performed percutaneously under sedation in the endoscopy suite[20]. Fixation to abdominal wall was obtained using also T-fasteners in a triangular disposition in the colon; the T-fasteners were cut at the skin after 4 wk. According to evidence, the experience of PES in pediatric settings is extremely limited, and this procedure should be reserved only for the small subset of children whit recurrent SV and high anesthesiological risks for open surgery.

***Is surgery necessary?***

After successful endoscopic reduction of the colon, the recurrence of SV was achieved in up two thirds of the cases. The largest data in adult population is provided by Atamanalp, who reported a 46-year experience with 952 patients with SV, in whom primary endoscopic derotation was successfully performed in 77% of patients, with the highest success rate in rigid sigmoidoscopy group (78.1%) compared with flexible sigmoidoscopy group (76.4%). A 4.5% of early recurrent rate was reported, and all the patients of this series eventually underwent elective or emergent surgical treatment[3]. In the pediatric review of Salas and colleagues, proctosigmoidoscopy and endoscopic rectal tube placement was attempted in 53.5% of cases, with a success rate of 47%[8]. Basing to the limited pediatric experience, we suggest that the initial endoscopic decompression and subsequent semi-elective operation results in a satisfactory outcome in managing SV. Waiting for surgery, a 48-72 h interval seems adequate for bowel preparation and optimization of the patient's clinical status[1,21]. Definitive semi-elective surgery is strongly recommend during the initial hospital admission for most of the patients[1]. Clinical evidence of peritonitis or perforation, unsuccessful endoscopic detorsion, gangrenous or ischemic bowel endoscopically evident obviously necessitates emergency surgical intervention[1,22].

***What are the risks of the endoscopic procedure?***

Inability to endoscopically detorse the SV is an indication for immediate surgical intervention. Shaft-induced perforations during endoscopy are due to a big loop formation. In these cases perforations are usually larger than expected and located on the antimesenteric wall. Tip perforations are smaller and typically occur when the “sliding by’’ technique is used inappropriately or a tip is trapped in wide diverticula or imbedded into mucosa when orientation is lost. Excessive air pressure perforation has been documented primarily in patients with strictures of the left colon, but are extremely uncommon in children[17,18]. In the historical review in adult setting provided by Atamanalp, iatrogenic perforations during endoscopy were recorded in 14 patients (2%); mortality rate of endoscopy was 0.05%[3]. Interestingly, no complications occurred during endoscopy were recorded in this review of pediatric series. To prevent excessive air insufflation water-immersion colonoscopy for SV was reported in adults[10-13].Nevertheless, according to evidence, the experience of water-immersion endoscopy in pediatric settings is extremely limited, especially in emergency setting.

**CONCLUSION**

Sigmoid volvuls in extremely uncommon in children and operative and technical details of endoscopic management is borrow by the larger adult experience. If no signs of bowel ischemia and perforation are present, water contrast enema followed by endoscopic decompression and detorsion of the volvulus represents the initial step of treatment also in pediatric setting. Nevertheless, the procedure requires a high degree of pediatric endoscopy expertise and is associated to high rate of early recurrence even when successfully performed. Elective surgery with sigmoid resection, primary anastomosis and sigmoidopexy is mandatory also in children successfully managed by endoscopic decompression and detorsion.

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**Table 1 Endoscopic management of sigmoid volvulus in childhood**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Ref** | **Number of patients** | **Demographic** | **Associated anomalies, *n* (%)** | **Endoscopic detorsion success rate, %** | **Recurrence rate, %** | **Surgery** |
| Salas *et al*[8] | 1 | M, 13 yr | irritable bowel syndrome | 100 | 100%, 2 days later | Sigmoidectmoy with colostomy and Harmann’s pouch |
| Salas *et al*[8] (Systematic review) | 63 | M:F =3.5:1, mean age 7 y | Hirschsprung’s disease: 11 (58%);  imperforate anus in 2 (11%); | 47 | 53% | Sigmoidectomy: 19 (38.7%);  Sigmoidopexy: 11 (22.4%)  Colostomy: 15 (30.6%) |
| Ton *et al*[7] | 1 | M, 16 y | - | 100 | 100% | Open sigmoid colectomy |
| Patel *et al*[2] | 1 | M, 14 y | Chronic constipation | 100 | 100% | Sigmoidectomy |
| Colinet *et al*[8] | 13 | M:F = 0.85:1, mean age 12.8 y | Mental retardation : 2 (15.3)  Myopathy: 2 (15.3);  Chronic intestinal pseudo-obstruction: 2 (15.3) | 100 | 50%, from 3 days to 3 months later | Sigmoidectomy 12 (84.6%) |
| Clermidi *et al*[6] | 1 | F, 11 y | Cornelia de Lange s. | 100 | 100 %, 2 days later | Open sigmoidectomy |
| Parolini *et al*[9] | 1 | F, 10 y | Functional constipation | 100 | 100 (%) | Sigmoidectomy and sigmoidopexy |

M: Male; F: Female.