Clinical Characteristics and Management of Colorectal Vascular Malformation in Children: A Retrospective Study of 23 Cases

Yanbing Huang[1,*] Peijun Zhuang[2,*] Gong Chen[1] Yanlei Huang[1] Kuiran Dong[1] Xianmin Xiao[1] Shan Zheng[1] Song Sun[1]

[1] Department of Surgery, Children's Hospital of Fudan University, Shanghai, China
[2] Department of Anesthesiology, Children's Hospital of Fudan University, Shanghai, China

Address for correspondence Song Sun, MD, Department of Surgery, Children's Hospital of Fudan University, Shanghai, China (e-mail: sun_song@fudan.edu.cn).

Eur J Pediatr Surg 2023;33:279–286.

Abstract

Introduction The objective of this study is to summarize the clinical characteristics and management of rare diseases of colorectal vascular malformation (CRVM) in children.

Methods We retrospectively analyzed the clinical data of CRVM patients admitted to the Children's Hospital of Fudan University from 2004 to 2019.

Results A total of 23 cases (16 males, 7 females) were enrolled. The median age of symptom onset was 1.4 years. Hematochezia and anemia were cardinal symptoms. Fourteen patients (60.9%) were misdiagnosed as anal fissures ($n = 4$), internal hemorrhoids ($n = 3$), rectal polyps ($n = 2$), inflammatory bowel disease ($n = 2$), portal hypertension ($n = 2$), and Meckel's diverticulum ($n = 1$), respectively. The average time from symptom onset to diagnosis was 4.5 ± 4.4 years. Other vascular malformations were detected in eight patients (34.8%). All patients showed a positive anomalous vascular image on contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI). The sensitivity of colonoscopy in the diagnosis of CRVM was 82.6% (19/23). A total of 21 patients underwent a modified Soave procedure. The lesions were mostly restricted to the colorectum and showed transmural diffuse distribution, with an average length of 20 ± 5.4 cm. Two patients (9.5%) experienced surgical complications. Bloody stools reappeared in two patients (9.5%), and colonoscopy showed abnormal angiogenesis at the anastomotic site, which were cured by sclerotherapy and/or electrocautery. The median follow-up time was 78 months. Bloody stools were absent at the last time of follow-up, and hemoglobin was in the normal range for all patients.

Conclusion The identification of CRVM in children often is delayed. Colonoscopy, CT, and MRI are essential in making the correct diagnosis. The modified Soave procedure is safe and feasible to treat CRVM in children. Endoscopic sclerotherapy and/or electrocautery can be used for residual lesions.

Keywords
- vascular malformation
- colorectal
- children
- hematochezia

[*] These authors contributed equally to this work and retain the first authorship.

received
April 5, 2022
accepted after revision
May 18, 2022
article published online
August 14, 2022

Georg Thieme Verlag KG,
Rüdigerstraße 14,
70469 Stuttgart, Germany

DOI https://doi.org/10.1055/s-0042-1751222.
ISSN 0939-7248.

Introduction

Colorectal vascular malformation (CRVM) is a rare, benign angiogenic lesion. CRVM is most often located in the rectosigmoid region, with primarily venous malformations (formerly known as "cavernous hemangioma").[1,2] The most common clinical symptom is recurrent, painless rectal bleeding that presents with melena, hematochezia, and anemia. Due to the nonspecificity of clinical presentation, CRVM is often misdiagnosed as hemorrhoids, colitis, polyps, or anorectal varices.[1–3] Endoscopic treatment, angiographic embolization, and surgical excision of the lesions have been reported to be effective in the treatment of CRVM.[4,5] However, due to the low incidence and unclear pathological mechanism, most of the previous studies were case reports, and there is still a lack of consensus on the diagnosis and treatment of CRVM in children. In this study, we reviewed 23 children with CRVM who were treated in our center from 2004 to 2019 to summarize experiences related to clinical characteristics and management.

Patients and Methods

We reviewed data from 23 patients treated for CRVM in the Department of Pediatric Surgery of Children's Hospital of Fudan University from 2004 to 2019. Patients with concomitant hemangiomas in the pelvic cavity, genitourinary tract, limbs, and skin were included. Hematochezia caused by simple hemorrhoid varices, portal hypertension, portal vein malformation, Abernethy malformation, inflammatory bowel disease, and other secondary colorectal varices or hyperplasia were excluded in this study. The clinical information that was collected included sex, age at symptom onset, age at diagnosis, the severity of rectal bleeding, misdiagnosis, treatment process, clinical outcome, and lesion distribution. We reviewed the consequences of assistant examinations, such as colonoscopy, computed tomography (CT), magnetic resonance imaging (MRI), and histopathology. Follow-up was conducted through face-to-face interviews in the outpatient department and telephone interviews. This study was approved by the Ethics Commission of Children's Hospital of Fudan University.

Table 1 Clinical characteristics of the 23 CRVM cases

Variables	$N = 23$
Gender, *n* (%)	
Male	16 (69.6)
Female	7 (30.4)
Age (y)	
Mean ± SD	2.8 ± 3.2
Median (range)	1.4 (0.1–12)
Time to diagnosis (y)	
Mean ± SD	7.1 ± 4.1
Median (range)	6 (1–16)
Symptoms, *n* (%)	
Hematochezia	23 (100)
Anemia	20 (87)
Abdominal pain	6 (26)
Other vascular malformation	8 (34.8)
hematuria/Vaginal bleeding	3 (13)
Hemoglobin (g/dL)	
Mean ± SD	75.8 ± 27.8
Median (range)	67 (39–120)
Blood transfusion, *n* (%)	
Yes	11 (47.8)
No	12 (52.2)
Misdiagnosis, *n* (%)	
Anal fissure	4 (17.4)
Hemorrhoids	3 (13)
Colitis	2 (8.7)
Varicosis	2 (8.7)
Polypus	2 (8.7)
Meckel's diverticulum	1 (4.3)
Location, *n* (%)	
Rectum	23 (100)
Sigmoid colon	20 (66.7)
Anal canal	4 (17)

Abbreviations: CRVM, colorectal vascular malformation; SD, standard deviation.

Results

General Situation

A total of 23 patients with CRVM were included in the study (16 males; 7 females). Basic patient information is listed in ►Table 1. The median age of bleeding onset was 1.4 years (range: 1 month–12 years) and most patients had been previously treated in hospitals. The median age at diagnosis was 6 years (range: 1–12 years) and the average time from symptom onset to diagnosis was 4.5 ± 4.4 years. Of the patients in the study, 23 (100%) had intermittent or persistent hematochezia, and 20 patients (87%) had anemia. The mean patient hemoglobin level was 75.8 ± 27.8 g/dL, and hemoglobin was lower than 90 g/dL in 17 patients (74%). A total of 11 patients (47.8%) had a history of repeated blood transfusion. Other symptoms included: mild lower abdominal pain in six patients, intermittent mild diarrhea in four patients, intermittent hematuria in two patients, varicose veins appearing at the anus during defecation in two patients, and occasional vaginal bleeding before puberty in one patient. Eight patients (34.8%) presented with vascular malformations in other locations. Two patients presented with skin vascular malformations of the trunk and extremities, accompanied by unilateral lower limb hypertrophy (►Fig. 1A). Other skin vascular malformations are distributed in the limbs, penis, and buttocks (►Fig. 1B–D). Abnormal vascular mass in the bladder or urethra was observed in two

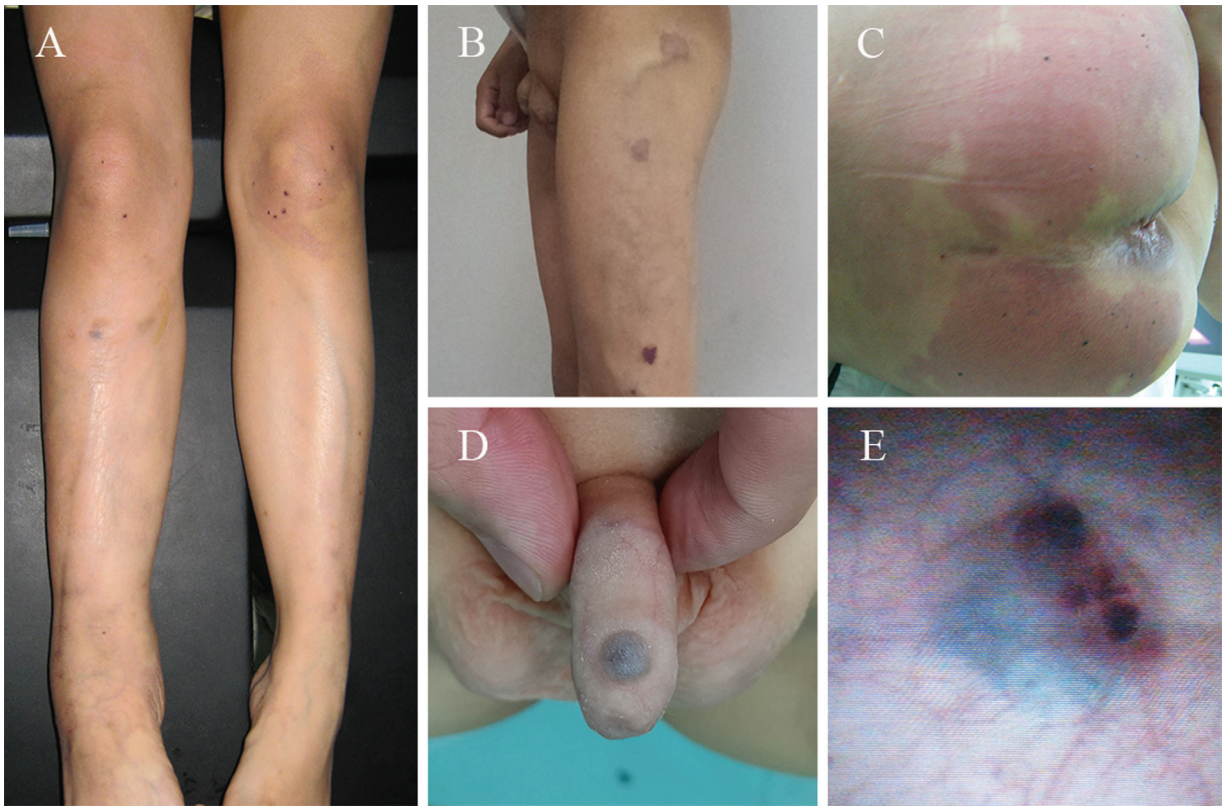

Fig. 1 (**A**) Asymmetry of lower extremities with the right leg demonstrating soft tissue and bony hypertrophy, as is typically seen in Klippel–Trenaunay syndrome (KTS). (**B**, **D**) Cutaneous vascular malformation of the lower limbs and perineum. (**C**) Port-wine stain on the buttocks. (**E**) Abnormal vascular mass in the bladder triangle.

patients (►**Fig. 1E**), and vaginal vascular malformation was observed in one patient.

Misdiagnosis and Mistreatment

A history of misdiagnosis in local hospitals was identified in 14 patients (60.9%). Four patients were misdiagnosed with anal fissures, and three patients were misdiagnosed with internal hemorrhoids before undergoing hemorrhoidectomy. Two patients were misdiagnosed with rectal polyps, one of which underwent endoscopic polypectomy. Two patients were misdiagnosed with inflammatory bowel disease and were treated with anti-inflammatory and hormone therapies. Two patients were misdiagnosed with portal hypertension, and rectal varices were treated with varicose vein ligation under a colonoscopy. One patient was misdiagnosed with Meckel's diverticulum and underwent laparoscopic exploration. Reduced bleeding was not achieved in any of the patients who received invasive treatment or conservative treatment.

Imaging Studies

All 23 patients were examined by colonoscopy. Lesions were identified to have originated from the dentate line and the mucosal lesions showed multiple focal distributions. The mean length of the involved bowel was found to be 14.3 ± 4.7 cm by colonoscopy. Abnormal submucosal vascular distribution could be seen by colonoscopy in 19 patients (82.6%; ►**Fig. 2**), and typically presented during colonoscopy as blue nodular lesions and dilated vessels in the mucosa or submucosa of the colon wall. The remaining four patients showed nonspecific manifestations, such as congestion, swelling, and bleeding (►**Fig. 2G, H**). The sensitivity of colonoscopy in the diagnosis of CRVM was 82.6% (19/23). A contrast-enhanced CT examination was performed on 17 patients (►**Fig. 3**). One or more positive signs, such as irregularly thickened rectal walls with varying degrees of enhancement, blurred perirectal spaces, or abnormal dilatation of blood vessels in or surrounding the rectum, were found on CT scans in all tested patients. Eight patients (46%) were found to have multiple calcification foci representing phleboliths (►**Fig. 3A**). Meanwhile, CT analysis also showed some concomitant diseases. One patient had a cystic space occupying the anterior portion of the right iliac vessel, which was considered a lymphatic malformation. Contrast-enhanced MRI was performed in 10 patients, which typically showed focal or multiple annular thickening of the rectal wall with low or equal signal intensity on T1-weighted images (T1WI), as well as significantly increased signal intensity on T2WI and fat-suppressed T2WI (►**Fig. 3D–F**). Moreover, MRI revealed the involvement of the pelvic cavity in two patients and the buttocks in another patient.

Treatment and Follow-Up

A total of 21 patients underwent a modified Soave procedure to remove the diseased intestinal segment; Sarasola–Klose hemorrhoidectomy was used for the distal portion of the endorectal dissection.[6] Three of these patients underwent a laparoscopy-assisted Soave procedure. Simultaneous loop ileostomy was

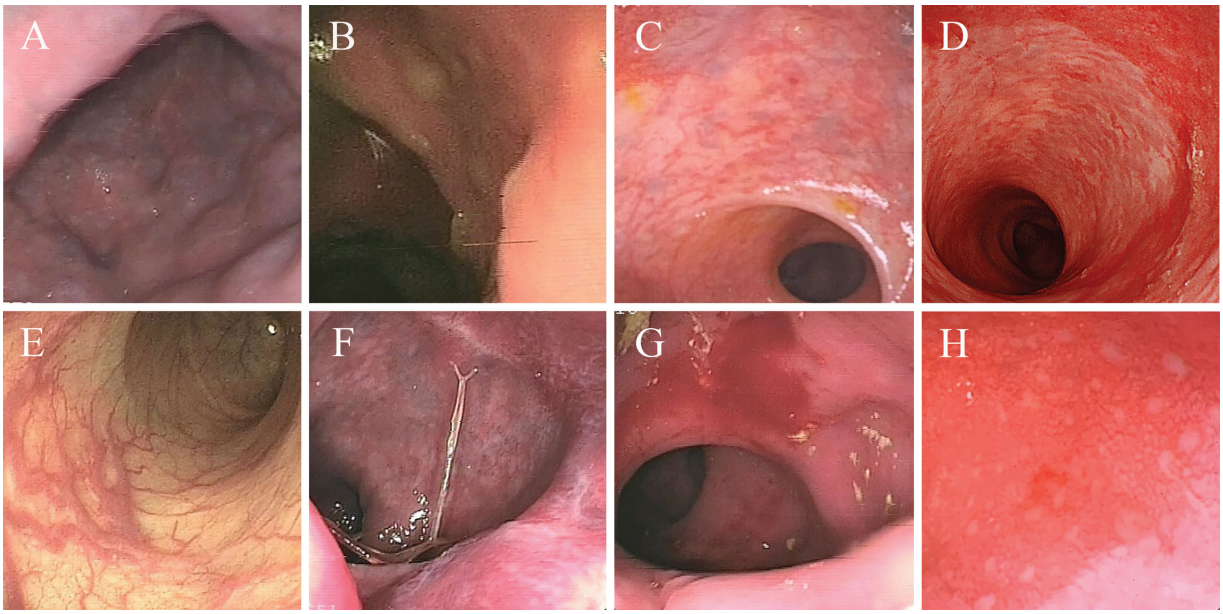

Fig. 2 Colonoscopic findings of colorectal vascular malformation (CRVM). (**A**) Serpentine dilated vessel. (**B**) Blue-purple nodules. (**C**) Sporadic blue patchy changes. (**D**) Mucosa ranging from bright red to bluish-purple. (**E**) Vascular thickening with prominent exposure. (**F**) Ischemic manifestations (scar-like changes). (**G**) Congestion, swelling, and bleeding. (**H**) White, round, spot-like changes.

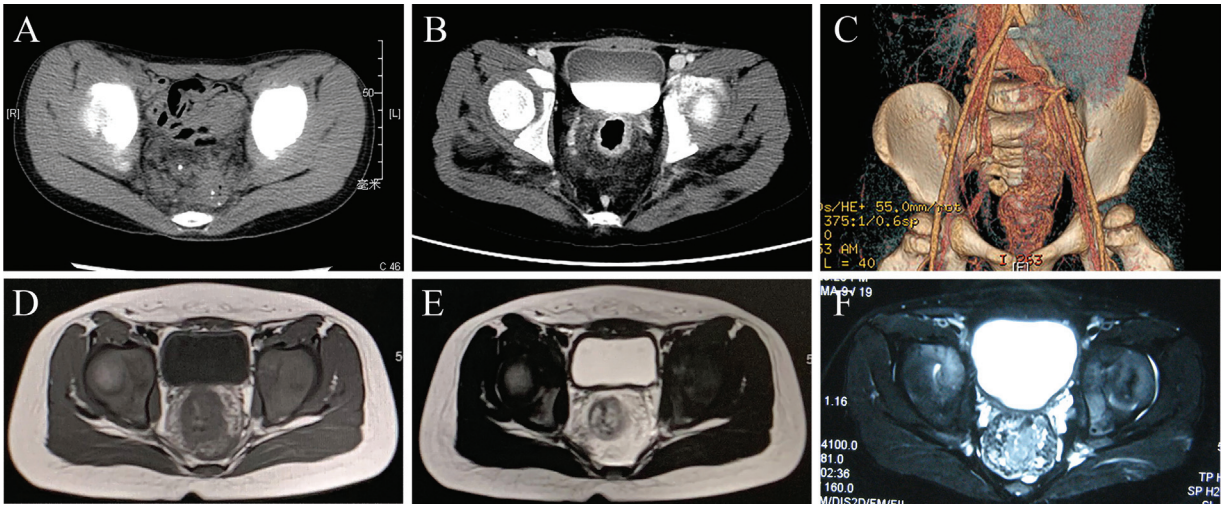

Fig. 3 Computed tomography (CT) and magnetic resonance imaging (MRI) findings of colorectal vascular malformation (CRVM). (**A**) Thickened rectal wall and multiple calcifications. (**B**) Circular enhancement of rectal mucosa. (**C**) CT angiography (CTA) shows more collateral branching of the venule around the rectum. (**D**) T1-weighted (T1WI) axial. (**E**) T2-weighted (T2WI) axial. The lesions were hypointense on T1WI and significantly hyperintense on T2WI. (**F**) A large number of tortuous cord-like signals in the mesorectum on the fat-suppressed (FS)-T2WI.

performed in 18 patients; the enterostomy was closed after a colonoscopy examination 3 to 6 months later. Two patients (9.5%) experienced surgical complications. One patient underwent a redo pull-through transection and anastomosis because of intestinal necrosis and anastomotic stricture. The other patient had reoperation of anastomotic stricture. Bloody stools reappeared in two patients, and colonoscopy showed abnormal angiogenesis near the anastomotic site, cured by sclerotherapy and/or electrocautery.

One patient diagnosed with Klippel–Trenaunay syndrome (KTS) presented with occasional slight rectal bleeding and was treated with conservative management and iron supplements. One patient with limited lesions underwent endoscopic foam sclerotherapy, and the patient was injected with lauromacrogol again after 6 months because of hematochezia recurrence.

Two patients with bladder vascular malformations who presented with focal eminence and erythema were subsequently cured by cystoscope electrocauterization. Urethral vascular malformations were not treated, and there was no recurrence of hematuria in the follow-up. The patient with massive vaginal bleeding achieved a significant remission after the Soave procedure, which may benefit from suture and ligation of the abnormal hyperplastic blood vessels around the uterus as much as is possible.

Five of the 23 patients failed to follow-up; the other 18 patients were followed for 6 to 184 months after surgery

(mean: 78 months). Two patients showed an increase in defecation frequency after enterostomy closure and were remitted with 6 months of conservative treatment. In three patients, intermittent fecal soiling was found within 3 months after the enterostomy closure. Two of these patients recovered gradually 3 to 8 months after surgery. All children had no bloody stool symptoms, and hemoglobin was in the normal range at the last time of follow-up.

Distribution of the CRVM

The lesions of 21 patients showed segmental diffuse distribution. The boundary of the normal intestine was clear, combined with the results of intraoperative exploration. Vascular tortuosity and dilatation could be seen on the serosa of the intestinal wall and thickened, and twisted blood vessels existed in some of the mesenteric vessels (►Fig. 4B–D). The lesions were mainly involved in the rectum, sigmoid colon, and descending colon; among them, two patients showed jumping distribution. In addition to rectal sigmoid colon involvement, ileum and transverse colon were also involved. Four patients were found to have anal canal involvement (►Fig. 4A). The average length of the involved intestine was 20 ± 5.3 cm, which was significantly longer than that of the lesions seen by colonoscopy (14.3 ± 4.7 cm, $p = 0.0158$). Upon histopathological examination, vascular malformations were shown to involve the whole intestinal wall in 11 patients, the submucosa and muscle layer in 7 patients, and the submucosa in 3 patients. Histopathology confirmed that primary vascular malformations were venous, and a few of them were complicated with lymphatic malformations (►Fig. 4E).

Discussion

CRVM is a rare gastrointestinal vascular malformation. Since CRVM was first described by Phillips in 1839, more than 300 cases have been reported, and most of these have been in adults.[7] Approximately 80% of CRVM lesions are located in the rectosigmoid.[1,2] The vascular malformation can present as solitary lesions, clusters of intraluminal polypoid lesions, or diffuse infiltration in the submucosa and other layers of the intestinal wall with extension into the mesentery and adjacent organs.[8] The classification and terminology of hemangioma and vascular malformations mentioned in the literature are not entirely clear and mainly include cavernous hemangiomas, angiodysplasia, arteriovenous malformation (AMV), venous malformation, and venous dilatation.[9,10] At present, CRVM is primarily reported to be a venous malformation (formerly called "cavernous hemangioma"), and AVMs are rare.[5,6,10] Previous studies have reported that CRVM affects more adult females than adult males,[11] but more male children than female children.[12] In our study, the results showed the morbidity of CRVM in male children was 69.6% (16/23), which is consistent with previous reports.

Chronic, recurrent, painless rectal bleeding is the most common symptom of CRVM. Occasionally, massive and life-threatening bleeding may occur.[1,6] Most patients often have

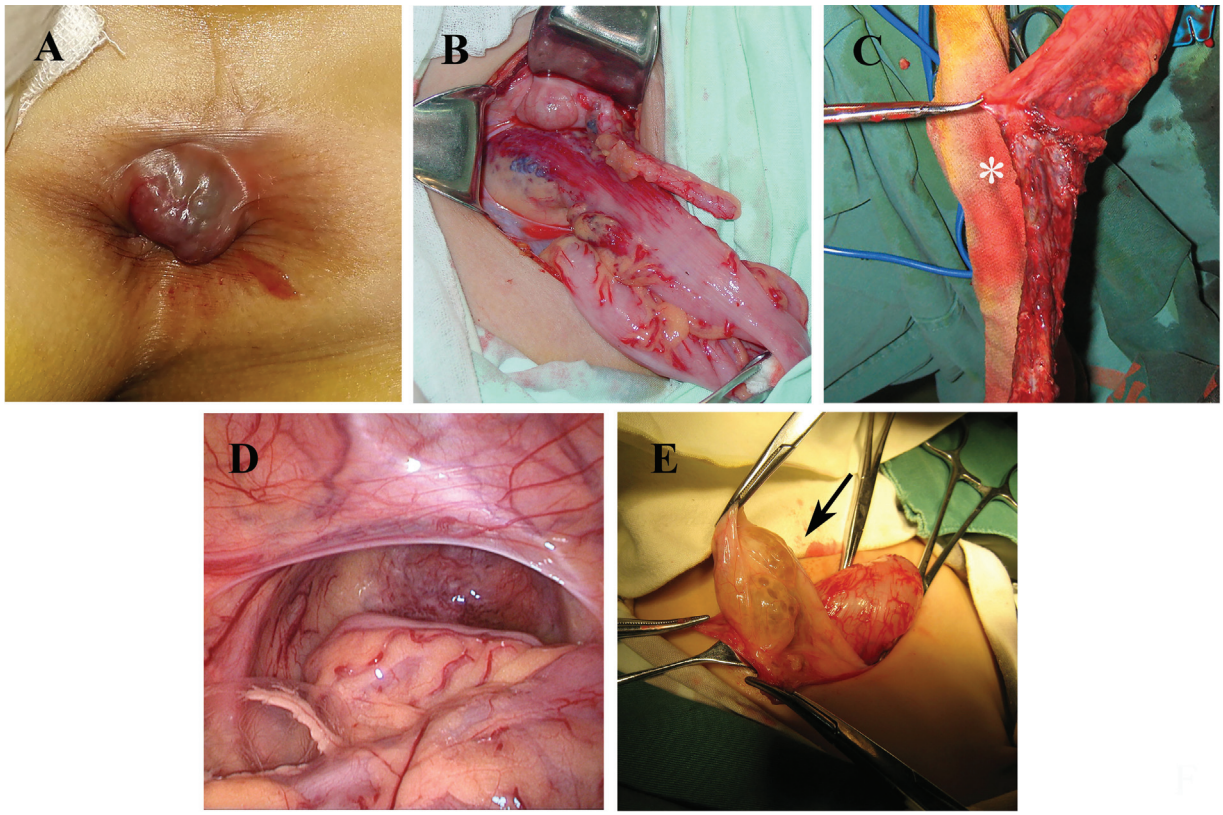

Fig. 4 **(A)** Vascular malformation involving the perianal region. **(B)** Vascular malformation was noted in the rectum and sigmoid colon. **(C)** Tortuous and enlarged blood vessels on the surface of the intestine. **(D)** Distribution of abnormal blood vessels in the pelvic cavity. **(E)** Lymphatic malformation.

a long-term history of repeated blood transfusions throughout the disease. Other reported but rare symptoms include abdominal discomfort, abdominal pain, diarrhea, and perianal discomfort.[2,8] Due to the rareness of CRVM and the lack of specific symptoms, a substantial number of patients have been misdiagnosed with anal fissures, hemorrhoids, rectal polyps, inflammatory bowel disease, and rectal varices. Wang et al reported that the mean delay time between the initial symptom onset and final CRVM diagnosis was 17.63 years, and the onset of CRVM in more than 50% of adults can be traced back to childhood or even infancy.[1,2] CRVM may manifest as intestinal lesions in syndromic diseases, such as KTS, which is characterized by CRVM, skin capillary malformation, and asymmetric limb hypertrophy.[13,14] In this study, eight patients (34.8%) had other associated vascular malformations, such as skin and genitourinary system, and two children were complicated with asymmetric hypertrophy of both lower limbs, which was considered KTS. Four patients had blood vessels dilate from the anal verge similar to hemorrhoids. Therefore, recurrent rectal bleeding, anemia, and hemorrhoid-like manifestations might be an indication of the CRVM in children, especially those who are associated with other vascular malformations.

Due to the low incidence of CRVM and the lack of large sample data, no consensus has been formed with respect to diagnostic methods. Colonoscopy is believed to be the most useful visual examination as it depicts the luminal extension and morphology of the malformation.[1,2,8] However, if the examiner is not experienced enough, atypical presentation by colonoscopy may lead to misdiagnosis. In the present series, 19 patients (82.6%) showed blue nodular lesions and dilated vessels in the mucosa or submucosa of the colon wall by colonoscopy. The other four patients showed atypical mucosal or submucosal lesions, such as congestion, swelling, and bleeding. We speculate that these features seen at the initial colonoscopy may have led to a diagnosis of colitis.

Contrast-enhanced CT is another important examination tool to confirm a CRVM. The representative findings of CT include an irregularly enhanced and thickened rectal wall or perirectal tissue, and multiple calcified foci of pelvic phleboliths.[1,2] Some of the CT findings of CRVM are nonspecific. However, when combined with CT angiography (CTA), the diagnostic efficiency can be greatly improved. CTA can clearly show even the smallest vessels of the malformation, as well as connections with other vessels of the circulatory system.[15] This has important guiding significance for intraoperative bleeding control. MRI has better tissue resolution than CT, which is of great significance in evaluating the extent of intestinal wall invasion and surrounding organ involvement and is important for guiding the choice of treatment.[15] Furthermore, dynamic contrast-enhanced MR angiography (MRA) can be used to differentiate between high- and low-flow lesions.[16] Typical imaging MRI findings show the rectosigmoid wall was markedly thickened with high signal intensity on T2WI, and an enhanced dynamic scan showed progressive enhancement. The addition of fat suppression to a fast spin-echo T2WI sequence is beneficial in depicting the true extent of the lesion, as it helps to differentiate the hyperintense lesion from the rectal mesentery. Digital subtraction angiography (DSA) can help to visualize the lesion and identify a vessel for embolization,[4] but it had a low positive rate of rectal vascular diagnosis (5/13, 38.5%) and could not show the lesion range.[2] With its low sensitivity, invasiveness, and high price, DSA can be replaced by CTA and MRA. Therefore, we suggest that enhanced CT and/or MRI are recommended as first-line examination methods for children with typical CRVM clinical manifestations.

Endoscopic ultrasonography has a good application prospect in the diagnosis of CRVM because it can accurately determine the level of intestinal wall involvement.[8,17] It can also evaluate the depth and direction of hemangiomas in the lower rectum and anal canal, especially for vascular abnormalities involving only the plasma layer, which cannot be recognized by colonoscopy. However, endoscopic ultrasonography has not been widely carried out in most children's endoscopic centers in China, and therefore, its application is limited.

CRVM treatments reported in the literature include surgical resection of the involved intestines, interventional embolization, and endoscopic treatment.[1,4,18] Surgical resection of the diseased intestinal segment is one of the more effective methods for treating CRVM. However, there is yet to be a consensus regarding specific surgical methods. The earliest reported surgical treatment was abdominoperineal resection, which involves the removal of a diseased intestinal segment to effectively stop bleeding.[2] Although this approach can save some patients with life-threatening severe intestinal bleeding, a permanent colostomy is undesirable, especially in young patients. Low anterior resection (LAR) can ultimately preserve sensation and defecation function of the anus[19]; however, LAR cannot completely remove the diseased rectum, and the probability of recurrent bleeding is very high. Therefore, this method can only be used for the treatment of vascular malformations above the sigmoid colon. In 1976, Jeffery first advocated for coloanal sleeve anastomosis for the treatment of CRVM, and this procedure has remained popular since.[20] However, this method is only able to remove the mucous layer and submucosa of the colorectum and retain a 10-cm-long rectal seromuscular layer. Therefore, vascular malformations of the seromuscular layer may invade the colon and cause bleeding recurrence. It is also tough to peel off 10 cm of the mucous membrane and seromuscular layer in the rectal wall with vascular malformations.

Recently, transanal total mesorectum excision for CRVM has been reported to contribute to the precise resection of the local lesion in the mid and low rectum and the anal canal with satisfactory outcomes when applied in the rectal cancer patients.[1,21] However, this operation completely removes the internal rectal sphincter and perirectal nerves, significantly influencing defecation sensation and defecation reflex functions after the operation. There is also a high risk of anastomotic leakage between a full-layer colon and low anal anastomosis.

Previous reports on surgical methods are mostly limited to adult cases. Due to the particularity of the anatomical structure of children, the adult procedure cannot be used

entirely. Previously, we used a modified transabdominal Soave procedure combined with Sarasola–Klose internal hemorrhoidectomy to treat CRVM, and achieved satisfactory results.[6] In this procedure, the laparotomy or laparoscopy was used to explore the extent of intestinal, mesenteric, and pelvic vascular malformations. The diseased mesentery is disconnected, and the colon and rectum are completely disintegrated below the peritoneal reflection, 3 to 5 cm proximal to the dentate line. The extent of disassociation can be determined by digital rectal examination during the operation. Turn to the perineum, and a gauze roll of 1.5 cm in diameter is slowly inserted into the rectum to a depth of about 3 to 5 cm. The distal end of the gauze roll and the rectal mucosa proximal to the dentate line are sutured. Circumferential incisions are made on the rectal mucosa's dentate line, and the gauze coils are pulled outward at the same time. Along the submucosal plane, the dissection is continued for another 3 to 5 cm proximal to the dentate line and enters the abdominal cavity. Then, the bowel is pulled down until the normal bowel appears. After the diseased bowel is resected, the cut edges of the colon are anastomosed to the dentate line. The internal sphincter and rectum muscle sheath are preserved, and the dissection range of the rectal mucosa is shortened in this procedure. Thus, the difficulty of the operation and the risk of bleeding are reduced greatly. A gauze roll is used to assist in achieving hemostasis by direct compression during dissection of the mucosa. The mucosa is fixed and retracted with tension, allowing the incision to be regular and the dissection plane to be well controlled. Due to the short distance from the peritoneal reflection to the dentate line in children, preserving a 3- to 5-cm rectal muscle sheath can significantly reduce perirectal nerve injury and minimize the effects on defecation.

In our center, 21 CRVM patients were treated with modified Soave surgery for complete resection of the diseased intestinal segment. Only two patients presented with abnormal vascular hyperplasia near the anastomotic site and hematochezia, which were cured by sclerotherapy and/or electrocautery. There was no reoperation due to the recurrence of hemangioma caused by the preservation of the muscle sheath. Therefore, even if the lesion begins from the dentate line, minor vascular malformations that might be retained in the bowel wall distal to the anastomosis after these resections can be easily managed by conservative treatment. Some children have a poor sense of defecation after the operation, likely due to the removal of the sensitive area of the mucous membrane on the dentate line, which requires slow recovery. Similar to the endorectal pull-through in the procedure for Hirschsprung's disease,[22] we noted an increased stool frequency in the early postoperative stage. This returned to normal by several months after the operation. Therefore, the modified transabdominal transanal Soave procedure is a suitable method for the treatment of CRVM because it can significantly reduce operation difficulty, bleeding risk, and postoperative defecation dysfunction.

In addition, our study found that mucosal or submucosal lesions of the intestinal wall identified by colonoscopy were significantly smaller than those of serous side lesions seen during operation. Children with long-segment lesions often showed patchy or small segment lesions by colonoscopy. Therefore, transabdominal surgery to explore the extent of the lesion is of great significance to prevent the omission of the diseased intestine and to prevent postoperative rebleeding. Pelvic floor vascular malformations around the peritoneal reflection can be seen during abdominal exploration in some children, which requires ligation around the rectum to control bleeding. In children with uterine and vaginal bleeding, the primary abnormal blood vessels around the uterus can be ligated during the operation to reduce vaginal or menstrual bleeding. Moreover, for a small number of children, abdominal lymphatic vascular malformations can be excised simultaneously with transabdominal exploration.

Nonoperative treatments for CRVM, such as sclerotherapy, snare polypectomy, neodymium-doped yttrium aluminum garnet laser therapy, and argon plasma coagulation, have been reported to decrease the amount and frequency of hematochezia.[23–25] However, rectal bleeding eventually recurred in most patients. In our center, one patient with limited lesions was treated with sclerotherapy and had recurrent hematochezia. The scope of rectal submucosal vascular malformation was gradually increased during follow-up. Moreover, sclerosing agent injection and interventional vascular embolization increased the risk of deep venous thrombosis, intestinal necrosis, pulmonary embolism, and even death. Data from several studies suggest that medical therapies are an effective and relatively safe treatment for patients with refractory bleeding from gastrointestinal vascular malformations.[11,26] Smith and colleagues reported that the patient was successfully managed using a conservative approach with tranexamic acid administered as needed, avoiding the need for resection.[7] However, the long-term efficacy of drugs in the treatment of CRVM remains inconclusive.

Funding

This study received financial support from the National Natural Science Foundation of China (nos. 81700450), Clinical Research Plan of SHDC (no. SHDC2020CR2009A), and Shanghai Municipal Key Clinical Specialty (no. shslczdzk05703). This study was approved by the Ethics Commission of Children's Hospital of Fudan University.

Conflict of Interest

None declared.

References

1 Zeng Z, Wu X, Chen J, Luo S, Hou Y, Kang L. Safety and feasibility of transanal endoscopic surgery for diffuse cavernous hemangioma of the rectum. Gastroenterol Res Pract 2019;2019:1732340
2 Wang HT, Gao XH, Fu CG, Wang L, Meng RG, Liu LJ. Diagnosis and treatment of diffuse cavernous hemangioma of the rectum: report of 17 cases. World J Surg 2010;34(10):2477–2486
3 Vitor S, Oliveira Ferreira A, Lopes J, Velosa J. Hemangioma of the rectum - how misleading can hematochezia be? Rev Esp Enferm Dig 2016;108(08):500–501
4 El-Sheikha J, Little MW, Bratby M. Rectal venous malformation treated by superior rectal artery embolization. Cardiovasc Intervent Radiol 2019;42(01):154–157

5 Fishman SJ, Burrows PE, Leichtner AM, Mulliken JB. Gastrointestinal manifestations of vascular anomalies in childhood: varied etiologies require multiple therapeutic modalities. J Pediatr Surg 1998;33(07):1163–1167
6 Lv Z, Xiao X, Zheng J, Liu J, Chen G. Modified Soave procedure for the treatment of vascular malformations involving anorectum and sigmoid colon. J Pediatr Surg 2009;44(12):2359–2363
7 Cotzias E, Rehman SF, Arsalani Zadeh R, Smith D. Conservative management of diffuse cavernous haemangioma of the sigmoid and rectum. Ann R Coll Surg Engl 2020;102(01):e1–e3
8 Li HB, Lv JF, Lu N, Lv ZS. Mechanical intestinal obstruction due to isolated diffuse venous malformations in the gastrointestinal tract: a case report and review of literature. World J Clin Cases 2020;8(01):157–167
9 Andrade P, Lopes S, Macedo G. Diffuse cavernous hemangioma of the rectum: case report and literature review. Int J Colorectal Dis 2015;30(09):1289–1290
10 Komekami Y, Konishi F, Makita K, et al. Rectal arterio-venous malformation (AVM) with bleeding of an internal hemorrhoid. Clin J Gastroenterol 2016;9(01):22–26
11 Ganesananthan S, Barlow J, Durai D, Hawthorne AB. Multiple venous malformations in the left colon and rectum: a long-standing case managed conservatively and an update of current literature. BMJ Case Rep 2019;12(03):e227700
12 de la Torre L, Carrasco D, Mora MA, Ramírez J, López S. Vascular malformations of the colon in children. J Pediatr Surg 2002;37 (12):1754–1757
13 Martinez-Lopez A, Salvador-Rodriguez L, Montero-Vilchez T, Molina-Leyva A, Tercedor-Sanchez J, Arias-Santiago S. Vascular malformations syndromes: an update. Curr Opin Pediatr 2019;31 (06):747–753
14 Masjedizadeh AR, Alavinejad P, Hashemi SJ. Rectal bleeding in a young patient with cavernous hemangioma and bone hypertrophy. Middle East J Dig Dis 2016;8(01):73–74
15 Bhattacharjee HK, Nariampalli Karthyarth M, Suhani S, et al. Laparoscopic total mesorectal excision for rectal venous malformation: a case report with a brief literature review. Asian J Endosc Surg 2021;14(01):85–89
16 Legiehn GM, Heran MK. Venous malformations: classification, development, diagnosis, and interventional radiologic management. Radiol Clin North Am 2008;46(03):545–597, vi
17 Sharma M, Adulqader A, Shifa R. Endoscopic ultrasound for cavernous hemangioma of rectum. Endosc Ultrasound 2014;3 (01):63–65
18 Markovic JN, Nag U, Shortell CK. Safety and efficacy of foam sclerotherapy for treatment of low-flow vascular malformations in children. J Vasc Surg Venous Lymphat Disord 2020;8(06): 1074–1082
19 Wang HT, Tu Y, Fu CG, et al. Diffuse cavernous hemangioma of the rectosigmoid colon. Tech Coloproctol 2005;9(02):145–148
20 Londono-Schimmer EE, Ritchie JK, Hawley PR. Coloanal sleeve anastomosis in the treatment of diffuse cavernous haemangioma of the rectum: long-term results. Br J Surg 1994;81(08): 1235–1237
21 Wu XR, Liang WW, Zhang XW, Kang L, Lan P. Transanal total mesorectal excision as a surgical procedure for diffuse cavernous hemangioma of the rectum: a case report. Int J Surg Case Rep 2017;39:164–167
22 Anupama B, Zheng S, Xiao X. Ten-year experience in the management of total colonic aganglionosis. J Pediatr Surg 2007;42(10): 1671–1676
23 Sierra A, Baixauli J, Elosúa T, Arredondo J, Cienfuegos JA. Life-threatening hematochezia from a rectal venous malformation successfully treated with microfoam sclerotherapy: description of a new technique. Int J Colorectal Dis 2015;30(10): 1427–1428
24 Gurien LA, Jackson RJ, Kiser MM, Richter GT. Nd:YAG laser therapy for rectal and vaginal venous malformations. Pediatr Surg Int 2017;33(08):887–891
25 Lee TY, Yeh HZ, Yang SS, Chang CS. Successful application of haemoclips plus argon plasma coagulation for angioectasia bleeding with underlying arteriovenous malformation in the colon. Colorectal Dis 2010;12(7 Online):e180–e181
26 Abematsu T, Okamoto Y, Nakagawa S, et al. Rectosigmoid colon venous malformation successfully treated with propranolol and celecoxib. J Pediatr Surg Case Rep 2015;3(08):331–333