

# Vascular Malformations of the Colon

Azam Ansari, MD

These lesions are believed to be the second most common cause of severe lower gastrointestinal bleeding, yet a lack of clinical awareness may cause delays in diagnosis and treatment.

**M**alformed or dysplastic components in the blood vessels that traverse the mucosal layers of the colon are known generally as vascular malformations of the colon (VMC). Such malformations may be innate or acquired and include vascular ectasias and some angiomas. They should not be confused, however, with hemangiomas, which are localized tumors of blood vessels expressing proliferation of cell nuclear antigen, vascular endothelial growth factor, and basic fibroblast growth factor.<sup>1</sup>

Although only 2% of all cases of lower gastrointestinal (GI) bleeding are attributable to VMC, these malformations may account for 15% to 30% of massive lower GI bleeding—which makes them second only to diverticulosis coli as a cause.<sup>2,3</sup> Historically, these lesions have been challenging to diagnose, especially during the acute bleeding episode. Since the advent of diagnostic angiography and colonoscopy in the clinical practice of gastroenterology, however, our knowledge of and techniques for managing VMC have improved. Nevertheless, there remains a paucity of information on this subject.

This article presents a brief review of the current state of VMC di-

agnosis and management, including discussions of disease classification, epidemiology, clinical presentation, essential diagnostic tools, and appropriate treatment options. The aim is to raise clinicians' awareness and expand their base of knowledge about this condition in order to promote prompt diagnosis and treatment and to encourage more investigation into this topic.

## PATHOPHYSIOLOGY AND EPIDEMIOLOGY

Because of imprecise and varied terminology, it is important to classify VMC on both an anatomic and clinicopathologic basis (Table 1). These two classification systems are not mutually exclusive. Anatomically, VMC can be arterial, venous, combined (containing both arterial and venous components), or capillary. Clinicopathologic classification

colon; 22% in the descending colon, sigmoid colon, or rectum; and 6% in the transverse colon.<sup>4</sup>

In symptomatic VMC, patients tend to be male with a mean age at diagnosis of 60 to 70 years. Population cross-studies have shown that 2% to 30% of individuals over age 50 may have VMC.<sup>5</sup> VMC also can occur in children<sup>6</sup>—which demonstrates that they are not exclusively degenerative lesions of aging.<sup>5</sup> In children, VMC usually are congenital.

Certain systemic diseases—such as chronic renal failure, radiation sickness, cirrhosis of the liver, aortic stenosis, and collagen vascular disease—may predispose patients to VMC. Recently, the causal association between aortic stenosis and VMC has been questioned,<sup>7</sup> and there is speculation that both conditions may occur independently as a result of age-related degeneration.<sup>8</sup>

*Frequently...the diagnosis is made incidentally during colonoscopy performed for other indications.*

Dr. Ansari is a gastroenterologist in the department of medicine at Fairview Southdale Hospital, Edina, MN. He is also a fellow of the American College of Gastroenterology.

considers the origin of the malformation: congenital, acquired, or inherited. Of all VMC, 33% occur in the cecum; 39% in the ascending

## CLINICAL PRESENTATION AND DIAGNOSIS

The majority of VMC are symptomatic, with the most common mode

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of presentation being intermittent or recurrent lower GI bleeding. In children with VMC, however, lower GI bleeding is less likely to be present, and intussusception or intestinal obstruction may be the only clinical manifestation.

In symptomatic patients, VMC may not be suspected until more common causes of lower GI bleeding are excluded. Frequently, in asymptomatic patients, the diagnosis is made incidentally during colonoscopy performed for other indications (such as colorectal cancer screening). The typically small size of VMC (generally, 2 to 20 mm) can add to the difficulty of diagnosis at initial presentation.

Various modalities are currently available for diagnosing VMC (Table 2), with endoscopic options being the most commonly used. In the absence of active bleeding, colonoscopy usually allows for ready identification of VMC, provided the clinician has a high index of suspicion and practices good mucosal cleansing (Figure).

During the active bleeding phase, even an experienced endoscopist may miss the diagnosis of VMC—unless spurting blood betrays the underlying lesion. Some of the difficulty of diagnosis during active bleeding may be mitigated through the use of a side viewing duodenoscope or through capsule endoscopy.<sup>9</sup> In addition, abdominal angiography and nuclear scintigraphy are alternative investigative modalities to consider, provided the rate of bleeding is 0.5 mL/min or higher. Visualization on angiography of minimal vascular tufts in the colonic wall or early venous filling will clinch the diagnosis.

In some cases, definitive diagnosis may not be possible without a histopathologic examination of a resected or autopsy-acquired specimen using

Table 1. Anatomic and clinicopathologic classification of vascular malformations of the colon	
Anatomic	Clinicopathologic
<p><b>Arterial</b></p> <ul style="list-style-type: none"> <li>• Ectasia</li> <li>• Angioma</li> <li>• Aneurysm</li> </ul> <p><b>Venous</b></p> <ul style="list-style-type: none"> <li>• Low flow                             <ul style="list-style-type: none"> <li>— Ectasia</li> </ul> </li> <li>• High pressure                             <ul style="list-style-type: none"> <li>— Varices</li> </ul> </li> </ul> <p><b>Combined</b></p> <ul style="list-style-type: none"> <li>• Arteriovenous malformation</li> </ul> <p><b>Capillary</b></p> <ul style="list-style-type: none"> <li>• Telangiectasia</li> </ul>	<p><b>Congenital</b></p> <ul style="list-style-type: none"> <li>• Seen in young patients, no predilection of site in the colon, involves all layers</li> </ul> <p><b>Acquired</b></p> <ul style="list-style-type: none"> <li>• Age-related, <i>without</i> underlying systemic disorder, in the right side of colon and limited to mucosa and submucosa</li> <li>• Age-related, <i>with</i> underlying systemic disorder (e.g., chronic renal failure, cirrhosis of liver, or aortic valve disease), limited to mucosa and submucosa</li> </ul> <p><b>Inherited</b></p> <ul style="list-style-type: none"> <li>• Autosomal-dominant, systemic disorder (Rendu-Osler-Weber disease)</li> </ul>

a silicone rubber compound injection technique.

be used in a complementary fashion (Table 3).

**MANAGING THE CONDITION**

Without definitive diagnosis and localization of the sites of VMC, management efforts can be frustrated by multiple relapses. The approach to treatment should be based, in part, on the specific type of VMC in order to improve prognosis and prevent relapse. Medical, endoscopic, and surgical modalities are available and may

**Medical treatment**

In select groups of patients (for example, those who have chronic renal failure or Rendu-Osler-Weber syndrome), oral estrogens have been known to prevent relapses, most likely by improving the integrity of the vascular endothelium and the coagulation components.<sup>10</sup> Iron supplements may be beneficial in those who

Table 2. Diagnostic modalities for vascular malformations of the colon	
<p><b>Noninvasive</b></p> <ul style="list-style-type: none"> <li>• Capsule endoscopy</li> </ul> <p><b>Semi-invasive</b></p> <ul style="list-style-type: none"> <li>• Colonoscopy</li> <li>• Nuclear scintigraphy</li> </ul>	<p><b>Invasive</b></p> <ul style="list-style-type: none"> <li>• Arteriography</li> <li>• Surgery</li> <li>• Histopathology*</li> <li>• Autopsy*</li> </ul>
*Involves silicone rubber compound injection of the specimen.	

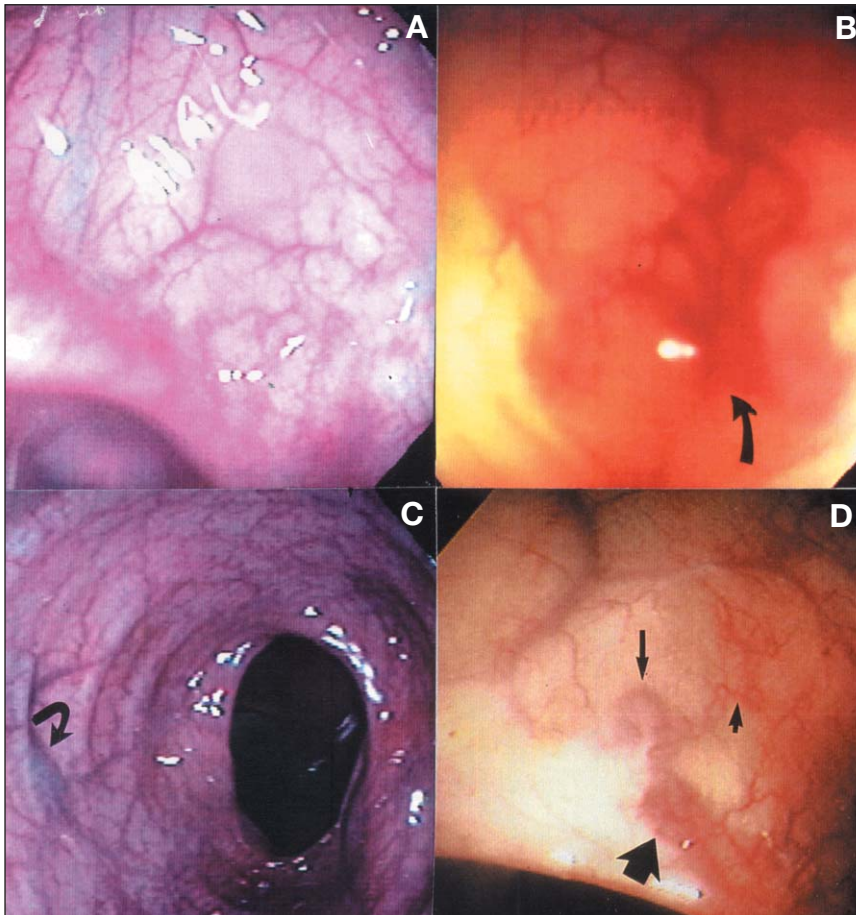


Figure. (A) Normal colon vasculature in a 52-year-old, white man seen during follow-up colonoscopy one year after polypectomy. (B) A barren, tree-like arterial angioma (curved arrow) found during colonoscopy in an 80-year-old, white man one year after removal of a villous adenoma. The arterial angioma was thermally ablated. There was no evidence of overt or occult gastrointestinal blood loss over one year. (C) Low-flow venous ectasia (bent arrow), shaped like a tennis racket, which was found during colonoscopy for colorectal cancer screening in a 54-year-old, white man. The malformation required no treatment. (D) A swan-shaped arteriovenous malformation (short, thick arrow), arising from an ectatic vein (long, thin arrow), and small arteries (short, thin arrow), which was found during colonoscopy for inflammatory bowel disease in a 72-year-old, white man. The malformation was thermally ablated, and there was no recurrence at two-year follow-up.

have a proven iron deficiency due to chronic intermittent blood loss.

## Endoscopic options

Currently, endoscopic treatment is the standard mode of VMC management. Heater probe application, bipolar or monopolar electrocoagulation,

ligating clip (Hemoclip, Teleflex Medical, Inc., Research Triangle Park, NC) application, sclerotherapy, and argon laser treatment all have their proponents.<sup>11,12</sup>

In the rare cases in which VMC present as a pedunculated polyp, excision by snare and electrocoagulation

leads to cessation of active bleeding.<sup>13,14</sup> More generally, one study found that endoscopic electrocoagulation resolves lower GI bleeding in 70% to 80% of VMC cases in which this type of bleeding is present.<sup>15</sup> In another study, 39% of such patients had relapses and 18% required surgery during long-term follow-up.<sup>16</sup>

## Surgical and interventional management

For patients with recurrent lower GI bleeding or numerous lesions, surgical or interventional treatment is indicated. Accurate identification of VMC sites is crucial before undertaking any interventional or surgical procedure in order to minimize morbidity and mortality.

Intra-arterial embolization is an interventional radiologic procedure that can be used to avoid surgery in some cases. It may be performed at the time of diagnostic arteriography.

At this time, the general consensus is that the use of blind right hemicolectomy in VMC is not tenable because of the high mortality rates (10% to 50%) resulting from concurrent cardiovascular and renal comorbidities.<sup>17</sup> Some authors, however, have justified subtotal colectomy as a last resort for unrelenting lower GI bleeding.<sup>17</sup> An alternative to this procedure, in cases of massive bleeding from VMC, is colectomy with mucosectomy and endorectal pull-through.<sup>18</sup>

## Follow-up care

Lower GI bleeding recurs in 37% of patients treated for VMC, even when other therapies are combined with surgery. In light of this fact, clinical and endoscopic follow-up is an important component of VMC management.<sup>19</sup>

## SUMMING UP

VMC are most common in the population over age 50. They are heteroge-

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**Table 3. Treatment modalities for vascular malformations of the colon**

<p><b>Medical</b></p> <ul style="list-style-type: none"> <li>• Estrogen</li> <li>• Iron supplementation</li> </ul> <p><b>Endoscopic</b></p> <ul style="list-style-type: none"> <li>• Thermal ablation                         <ul style="list-style-type: none"> <li>— Heater probe application</li> <li>— Bipolar electrocoagulation</li> </ul> </li> <li>• Ligating clip application</li> <li>• Sclerotherapy</li> <li>• Argon laser application</li> </ul>	<p><b>Interventional</b></p> <ul style="list-style-type: none"> <li>• Vascular obliteration by intravascular embolization</li> </ul> <p><b>Surgical</b></p> <ul style="list-style-type: none"> <li>• Segmental colectomy</li> <li>• Colotomy with mucosectomy and endorectal pull-through</li> </ul>
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neous in their appearance and mode of presentation. Previously linked with aortic stenosis, most VMC are now believed to be age-related vascular lesions. Anatomic and clinicopathologic classifications of VMC have been developed to reduce confusion over varied terminology. Colonoscopy plays an important role in the diagnosis and management of this condition. Endoscopic treatment is the preferred approach. Arterial embolization or surgery is indicated when bleeding continues or recurs after endoscopic treatment. ●

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