

# Endovascular and Percutaneous Treatment of Pediatric Portosystemic Varices: A Case Review

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## Abstract

Portal hypertension is a significant cause of morbidity and mortality in pediatric patients. Complications of portal hypertension include development of portosystemic varices. The most common type of portosystemic varices are gastroesophageal varices; however, other ectopic varices can also be a cause of recurrent, life-threatening gastrointestinal bleeding. Problematic ectopic varices include isolated gastric, anorectal, small bowel, roux-limb, and stomal varices. There are no standardized treatment guidelines on how to manage ectopic varices in children; however, new innovations in endovascular treatment options provide potential therapeutic alternatives when varices are refractory to conventional therapy. This review provides a case-based literature review for endovascular treatment of isolated gastric, anorectal, small bowel, roux-limb, and stomal ectopic varices in children (age 0-9 years) and adolescents (age 10-19 years).

## Keywords

- ▶ portosystemic varices
- ▶ ectopic varices
- ▶ venous intervention
- ▶ pediatric interventions

Portal hypertension in children is a significant cause of morbidity and mortality. One common complication associated with portal hypertension is the development of portosystemic varices, such as gastroesophageal and other ectopic varices (EcV). Portal hypertension in adults is defined as a hepatic venous pressure gradient greater than 5 mm Hg, and variceal formation occurs from a hepatic venous pressure gradient  $\geq 10$  mm Hg leading to venous collateralization, classically in predictable locations.<sup>1</sup> The most common type of varices are esophageal and gastroesophageal varices. However, less-common, yet equally as problematic, EcV include isolated gastric, anorectal, small bowel (most commonly duodenal), roux-limb, and stomal varices (▶ **Figs. 1** and **2**). Acute life-threatening variceal bleeding is the most

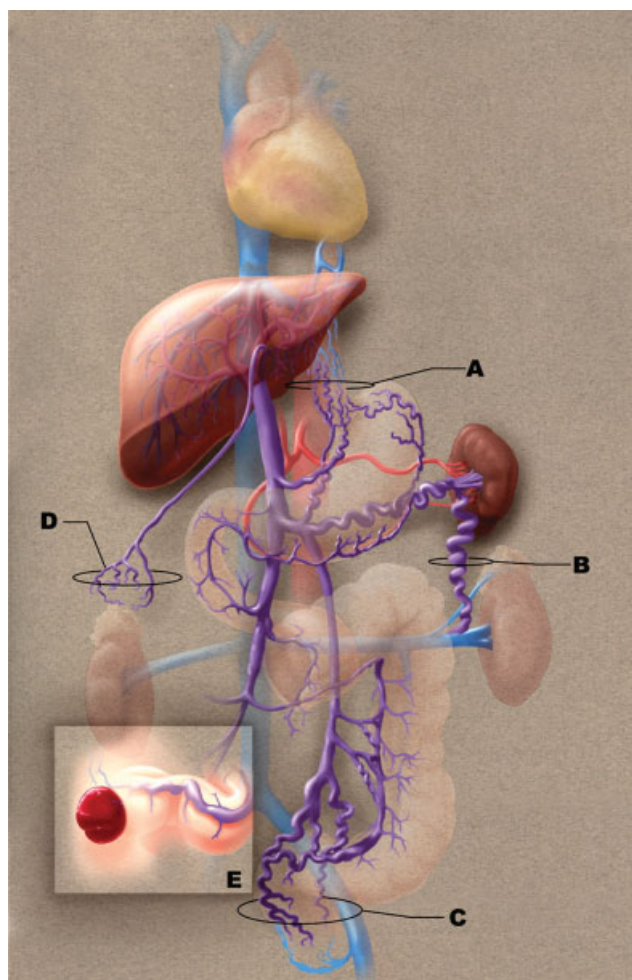
feared complication of all types of EcV. Traditionally, in children, most EcV have been treated both endoscopically and surgically. However, minimally invasive percutaneous and endovascular therapies have evolved as potential therapeutic options to treat EcV not amenable to endoscopic or surgical treatment in children. Transjugular intrahepatic portosystemic shunt (TIPS) creation has been extensively described in the peer-review literature to treat pediatric esophageal and gastroesophageal varices,<sup>2-5</sup> and will, thus, not be extensively reviewed herein. This review, rather, focuses on the literature to date in a case-based and location-based approach to percutaneous and/or endovascular treatment of pediatric isolated gastric, anorectal, small bowel, roux-limb, and stomal EcV.

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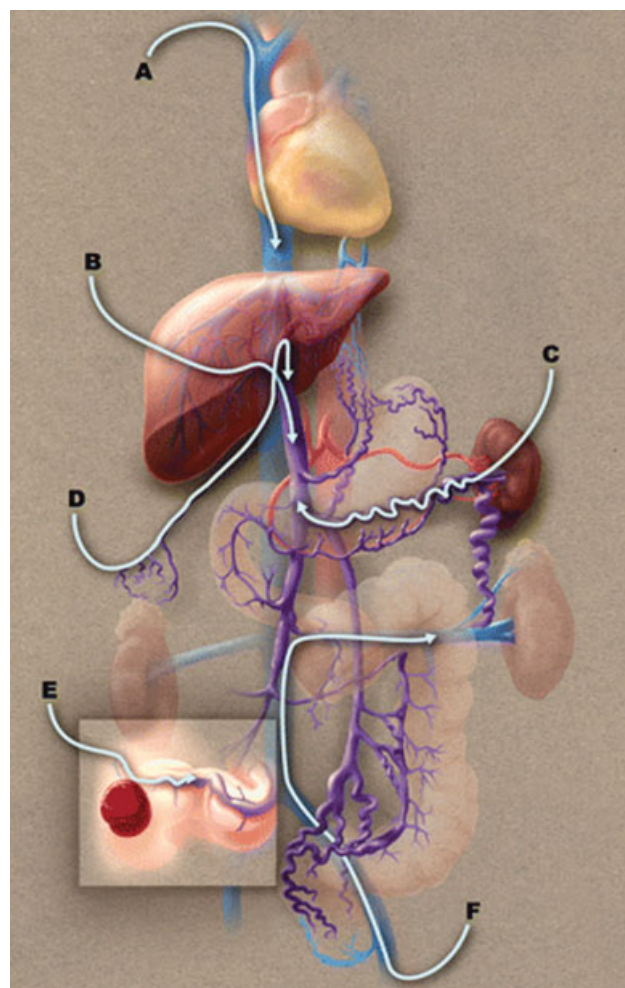
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**Fig. 1** Drawing illustrates the common portosystemic venous collateral pathways: the left gastric (coronary) and azygos veins at the lower esophagus (a); the splenic and left renal-adrenal veins at the spleen (b); the superior rectal and middle-inferior rectal veins at the anal canal (c); the veins of the ligamentum teres and the superior-inferior epigastric veins at the umbilicus (d); and the colonic-mesenteric veins, body wall veins, and IVC in the retroperitoneum (e). (Referenced from Glen Oomen, MSc, McMaster University, Dundas, Ontario, Canada.)

The pediatric literature is limited in its description of the risk factors for developing EcV, as invasive hemodynamic measures are rarely performed and thus inferences are made from the adult literature including using similar hepatic vein pressure gradient thresholds as adults.<sup>6</sup> In adults, there is a direct correlation between the degree of hepatic cirrhosis measured by Child Pugh or Model for End-Stage Liver Disease (MELD) scores and degree of hyperdynamic circulation and portal pressure gradient.<sup>7</sup> Up to 60% of patients with decompensated cirrhosis (as evidenced by coagulopathy, ascites, and hepatic encephalopathy) and up to 30% of patients with compensated cirrhosis have varices. Approximately 10% to 20% of these patients' varices increase in size within 1 to 2 years. Importantly, size of esophageal varices is directly correlated with risk of variceal bleeding<sup>8</sup> and variceal size is directly related to the degree of hepatic vein wedge pressure gradient elevation. Significant acute variceal bleeding is associated with up to 50% mortality<sup>9</sup> in adults, and, therefore, close attention to patients with EcV is necessary.



**Fig. 2** Drawing illustrates the various routes that can be used to access the portal venous system in complicated portal hypertension, including transjugular (a), transhepatic (b), trans-splenic (c), trans-umbilical (d), transvariceal (stomal) (e), and transfemoral (systemic-portal) (f) routes. (Referenced from Glen Oomen, MSc, McMaster University, Dundas, Ontario, Canada.<sup>94</sup>)

However, more recent literature demonstrates pediatric mortality from variceal bleeding is uncommon,<sup>10</sup> although long-term morbidity in these patients is not well described.<sup>6</sup> Often times, gastrointestinal bleeding from a ruptured varix is the most common presenting symptom of portal hypertension. Life-threatening bleeding and complications occur mainly in patients with portal hypertension secondary to hepatic cirrhosis due to the downstream complications of hepatic decompensation.

### Pediatric Etiologies

There are fundamental differences in the pathophysiology resulting in portal hypertension and subsequent EcV in pediatric patients when compared to adults. There are many different causes of pediatric portal hypertension, which can further be subdivided into extrahepatic (prehepatic and infrahepatic), intrahepatic, and post hepatic (► Table 1).

**Table 1** Causes of pediatric portal hypertension<sup>91–93</sup>

Intrahepatic	Biliary disease	Biliary atresia Progressive familial intrahepatic cholestasis Primary sclerosing cholangitis Cystic fibrosis Intestinal failure-associated liver disease
	Hepatic disease	Autoimmune hepatitis Chronic viral hepatitis B/C Alpha <sub>1</sub> -Antitrypsin deficiency Nonalcoholic fatty liver disease Wilson's disease Glycogen storage disease
	Fibrotic disease	Congenital hepatic fibrosis Caroli disease
Prehepatic	Portal vein occlusion	Portal vein thrombosis Tumor infiltration or compression Congenital atresia/agenesis
	Splenic vein thrombosis	
	Increased portal flow	Arteriovenous fistula
	Nodular regenerative	Drug therapy (6-thioguanine and azathioprine) Turner syndrome
	Portal stenosis	Schistosomiasis Idiopathic cause HIV infection Cystic fibrosis liver disease
Posthepatic	Hepatic vein obstruction	Budd–Chiari syndrome Inferior vena cava obstruction Congestive heart failure Veno-occlusive disease (sinusoidal obstruction)
	Cardiac disease	Right-sided heart failure Cardiac tamponade

Although portal hypertension is mainly secondary to hepatic cirrhosis in adults, the two most common causes of portal hypertension in children are cirrhosis, with biliary atresia as the leading cause in developed countries<sup>11</sup> and extrahepatic portal vein occlusion in developing countries and second most common cause of portal hypertension in developed countries.<sup>12–14</sup> The discrepancy between developed and developing countries is not entirely clear, but the recorded prevalence of hepatitis B virus and Wilson's disease is higher in similar cohorts in developing countries which suggests underdiagnosis and/or undertreatment of these two conditions.<sup>11,12</sup> In children who have received a liver transplant, studies have demonstrated portal venous thrombosis and stenosis rates of 3.7% to 50% in pediatric patients, irrespective of the reason for liver transplant,<sup>15–17</sup> which can form new EcV or worsen existing EcV. Interestingly, post-transplantation portal venous thrombosis or stenosis is independently associated with increased mortality.<sup>17</sup>

Serial endoscopy for surveillance of gastroesophageal varices is discouraged in children, and noninvasive, highly accurate

clinical prediction rules have been created based on spleen size, hypoalbuminemia, and platelet count to predict risk of forming esophageal varices.<sup>11,18</sup> However, no such prediction rules have been delineated for the less common types of EcV discussed in this review likely due to the lack of published treatment and outcomes data.

### Baveno Consensus Recommendations: Prophylaxis versus Acute Variceal Bleeding

Beginning in 1990, in response to the scarce literature on approaches to management of pediatric variceal bleeding, an expert panel has convened every 5 years in Baveno, Italy, to explore the data in pediatric variceal bleeding. The most recent consortium of the “Controversies in the Management of Varices in Children—An International Forum” met in 2015, and from this meeting, experts recommend no specific primary prophylaxis with either medication (nonselective beta-blockers), endoscopic variceal ligation, or sclerotherapy based on the lack of evidence. Meso-Rex Bypass (a vascular graft between the superior mesenteric vein and left portal vein) may be beneficial for primary and secondary prophylaxis in certain cases of extrahepatic portal vein occlusion.<sup>19</sup> Endoscopic ligation and sclerotherapy can be performed to achieve hemostasis in gastroesophageal varices. In addition, for acute variceal bleeding, octreotide has been shown to be safe and effective in the majority of cases.<sup>20</sup>

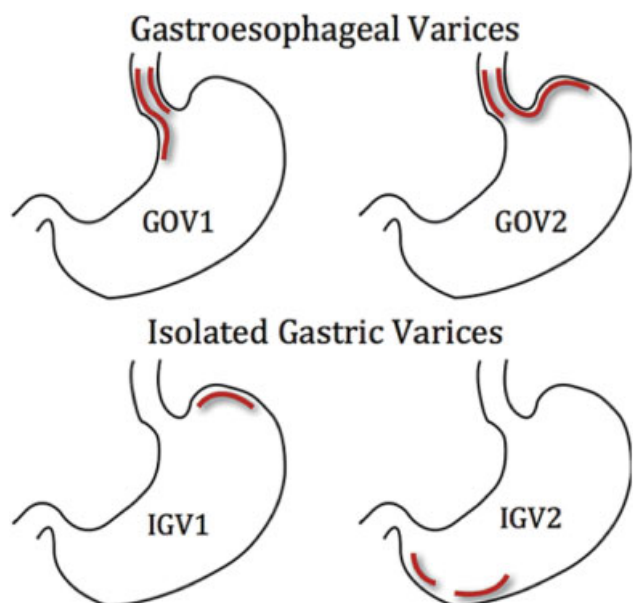
Within the newest 2015 Baveno recommendations, endovascular therapy, specifically TIPS, was not directly addressed. However, the recommendations from the 2010 Baveno forum suggested that early TIPS placement (within 72 hours, ideally 24 hours) can be considered in the setting of chronic liver disease in patients with bleeding EcV that have failed initial pharmacological and endoscopic therapy.<sup>21</sup>

### Endovascular Therapy for Different Types of Ectopic Varices

#### Gastroesophageal Varices

Gastroesophageal varices are the most common types of varices that occur in patients with portal hypertension. Although prevalence rates are not well defined in the pediatric literature, in adults, approximately 50% of patients with cirrhosis develop gastroesophageal varices and these varices are present in 5% to 33% of patients with portal hypertension. The rates of developing varices is approximately 5% to 8% annually with 1% to 2% of patients developing large enough varices placing them at risk for bleeding.<sup>22</sup> Gastroesophageal varices have four classifications: gastroesophageal varices (GOV) 1 and 2 and isolated gastric varices (IGV) 1 and 2 (►Fig. 3). Importantly, recent literature suggests that TIPS is insufficient in treating isolated gastric varices with gastric variceal post-TIPS patency of 65%, and recurrent bleeding rates of 27%,<sup>23</sup> suggesting more extensive occlusion of these varices is required for improved outcomes.

TIPS been researched extensively for decades in adults and is a safe and efficacious treatment option for gastroesophageal varices.<sup>24</sup> More recently, retrospective reviews demonstrate



**Fig. 3** Types of gastroesophageal (GOV) and isolated gastric varices (IGV). (Adapted from Sarin et al.<sup>95</sup>)

TIPS shunts placed in pediatric patients demonstrated efficacy and safety comparable to that published in adults.<sup>2,5</sup> Furthermore, a thorough review of pediatric TIPS was recently published by Monroe et al in February 2018.<sup>3</sup> Thus, despite TIPS being the most commonly performed endovascular procedure for treatment of bleeding esophageal and gastroesophageal varices in children, the technical specifics of TIPS placement will not be discussed further in this manuscript.

### Isolated Gastric Varices

Balloon-occluded retrograde transvenous obliteration (BRTO) has emerged as a safe, and potentially efficacious, minimally-invasive therapeutic option for treating isolated gastric varices in certain patients, particularly IGV2. BRTO was first described in adults in Japan in the early 1990s, and the first case reports of pediatric BRTO were described in the early 2000s. To date, case reports of pediatric BRTO have been published on pediatric patients from 2 to 15 years of age.<sup>25–31</sup> The most commonly described technique in the literature to access the bleeding gastric varix is through a patent portocaval shunt as gastric varices drain by a gastrosplenorenal (87%) shunt or a gastric-inferior phrenic (16%) shunt.<sup>32</sup> If the gastrosplenorenal shunt is large enough to facilitate a retrograde approach, BRTO may be an appropriate treatment (► Fig. 4a–c). Successful pediatric BRTO has been reported in gastric varices secondary to biliary atresia, extrahepatic portal venous obstruction, and post-liver transplant portal hypertension<sup>26,29,31,33</sup> in addition to congenital portosystemic (superior mesenteric vein–inferior vena cava and splenorenal) shunts.<sup>27</sup> Alternatively, a trans-splenic approach may be used to access the varices if no large, spontaneous portosystemic shunt has developed (► Fig. 5a–g). Ultimately, pediatric BRTO can be successful in reducing the size and tension of gastric varices.

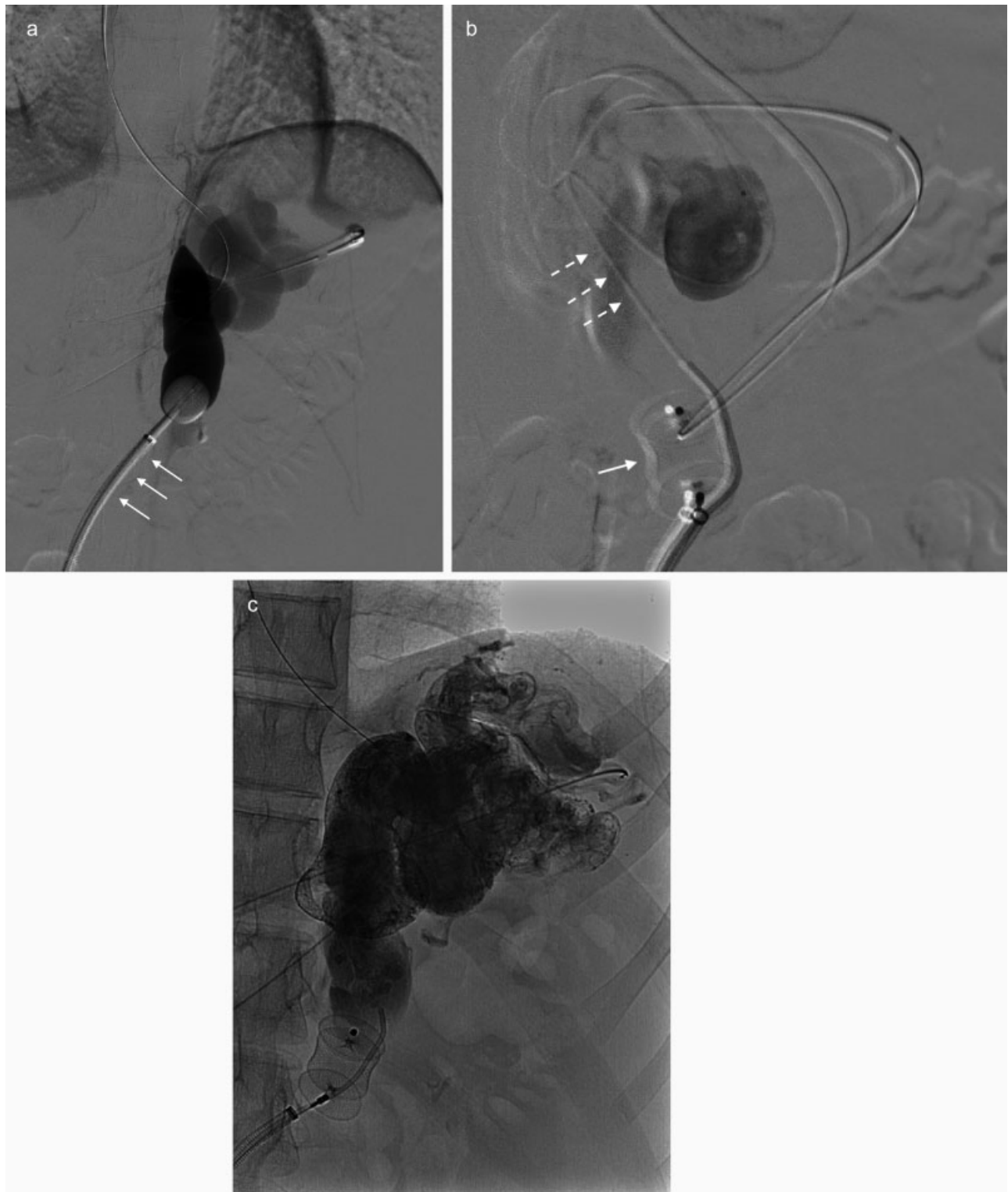
More recent literature demonstrates an alternative method to improve filling of the gastric varices during BRTO with

temporary balloon-occlusion of the splenic artery.<sup>34</sup> Temporary occlusion of the splenic artery with a 5-F balloon reduces pressures in the vein draining the gastric varix which adequately fills the gastric varix with the sclerosing agent, resulting in improved varix obliteration.<sup>35</sup>

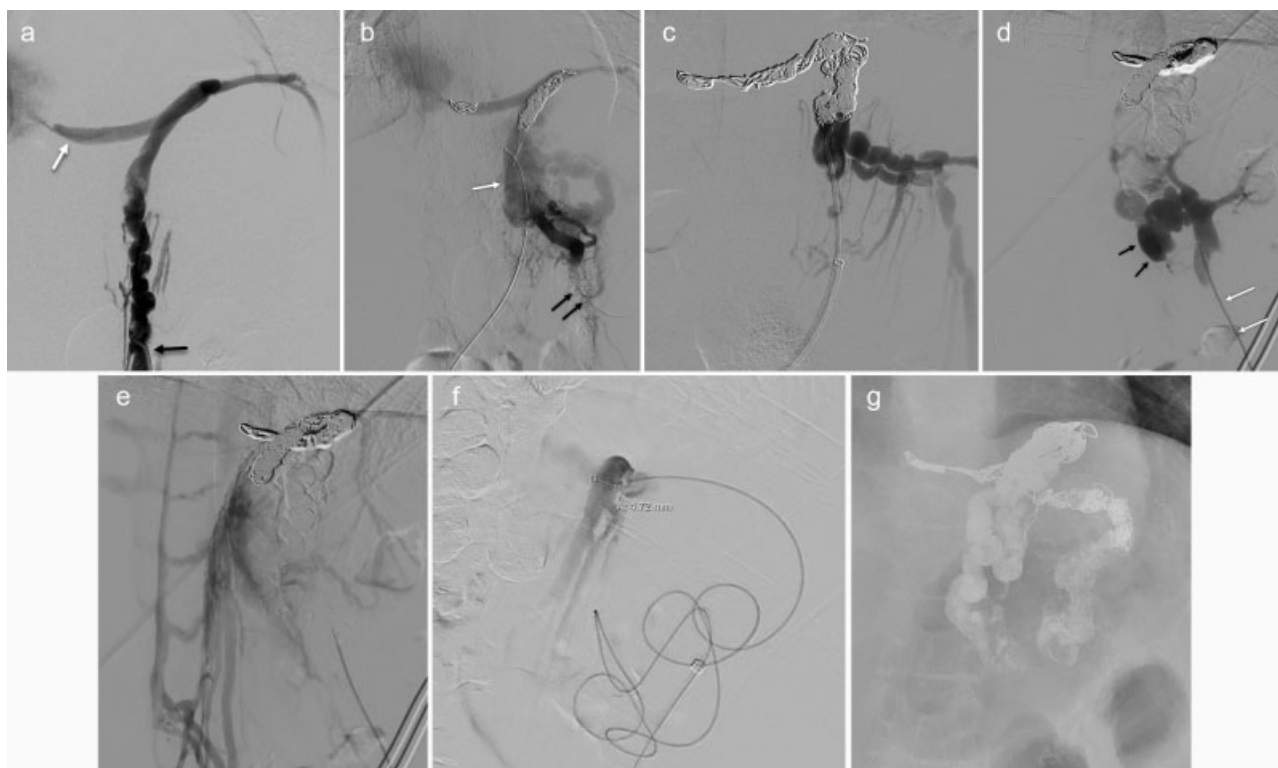
The most common sclerosing agent used in the Japanese literature is ethanolamine oleate (Oldamin, Aska Pharmaceutical, Tokyo, Japan). However, there is a risk of hemoglobinuria-related acute renal failure secondary to hemolysis from ethanolamine oleate,<sup>36,37</sup> and prophylactic haptoglobin (Benesis, Osaka, Japan) can be administered preoperatively<sup>27,38</sup> to reduce this risk. Since haptoglobin is not available in the United States, alternative sclerosing agents and embolics with a lower risk of hemolysis are typically used, such as sodium tetradecyl sulfate (STS) (Sotradecol, AngioDynamics, Inc., Queensbury, New York), polidocanol (Polidocasklerol, ZERIA Pharmaceutical, Tokyo, Japan), *n*-butyl-2-cyanoacrylate (NBCA, Trufill, Cordis Neurovascular), and 50% glucose.<sup>39</sup> A foam sclerosant can be created with air or carbon dioxide to increase the injected volume without increasing the dosage of sclerosant, ensuring adequate contact with the endothelial cells of the varix. Common combinations in the United States include 1 part ethiodized oil (Lipiodol, Ethiodol, Savage Laboratories, Melville, New York), 2 parts 3% Sotradecol, and 3 parts carbon dioxide,<sup>39</sup> or air. At our institution, our pediatric maximum dosage of 3% STS is 20 mL (0.5 mL/kg) and ethiodized oil is 0.25 mL/kg for all sclerosant mixtures.

### BRTO Outcomes

Overall, BRTO is a well tolerated procedure with up to 95.5% success rate in a sample of 154 adult patients with similar results in a much smaller cohort of published pediatric patients.<sup>38</sup> Causative portal hypertension, however, may be exacerbated by the loss of decompressive spontaneous shunts emphasizing the importance of patient selection prior to BRTO. The obliteration of the spontaneous hepatofugal shunts in these patients can increase portal hypertension, worsening esophageal varices and ascites.<sup>40</sup> Esophageal varices worsening following BRTO has been recorded in 10% to 63% of patients, with more recent literature demonstrating longitudinal exacerbation of esophageal varices at 1, 3, 5, and 7 years of 13%, 20%, 27%, and 35%, respectively. Overall 5-year survival rates after BRTO are 50% to 72% in the adult populations.<sup>38</sup> In adults, immediate postoperative clinical complications include transient fever (33%), chest or epigastric pain (56%), hemoglobinuria (49%), hypertension (35%), nausea or vomiting (21%), gastric ulcers (9%), and hemorrhagic portal hypertensive gastropathy (2%). Less common complications (7 to 10 days postoperatively) include: pleural effusion (12%), and pulmonary infarction (2%).<sup>41–43</sup> Routine monitoring of laboratory values one day post-BRTO demonstrate significant increases in indirect bilirubin, aspartate aminotransferase, and lactate dehydrogenase, returning to normal pre-BRTO levels by day seven.<sup>42</sup> Total bilirubin significantly increases one day post-BRTO; however, can be elevated for upwards of 36 months post-BRTO particularly in the setting of a patent splenorenal or gastrosplenic shunt.<sup>41,42</sup>



**Fig. 4a–c** A 16-year-old girl with biliary atresia status post Kasai hepatopertoenterostomy procedure with isolated gastric varices originally noted on endoscopy presented with severe hematemesis and a hemoglobin of 4 g/dL and was nonresponsive to transfusions. ► **Figure 4a** shows opacification of large gastric varices following access into a gastrosplenorenal shunt from the right femoral vein and left renal vein with a 9F sheath (white arrows) and a 5F Python balloon-occlusion catheter (5F is the maximum occlusion balloon size for pediatrics). Through the sheath, a 10mm AVP2 Amplatzer (St. Jude, Plymouth, Minnesota) occlusion device (► **Fig. 4b**, white arrow) was deployed (but not detached). Alongside the occlusion device, a 4F angled catheter (Terumo, Somerset, New Jersey) (black arrow) was advanced, through which a 2.5F Cantata (Cook Medical Inc, Bloomington, Indiana) microcatheter (dashed white arrows) and 0.014 Synchro microwire (Stryker, Kalamazoo, Michigan) were used to obtain deep access into the gastric varices. ► **Figure 4c** shows the distribution of 3% Sotradecol (AngioDynamics, Inc., Queensbury, New York) embolic foam (3 parts air: 2 parts 3% STS:1-part ethiodized oil, Guerbet, Princeton, New Jersey) throughout the varices. The AVP2 occlusion device was then deployed without egress of the embolic foam. The patient's bleeding and hematemesis ceased immediately. She received a transplant liver shortly thereafter.



**Fig. 5a–g** An 18-year-old male with autoimmune hepatitis resulting in severe portal hypertension demonstrating large isolated gastric varices, anemia, and melena. Injection from a gastrorenal shunt (►Fig. 5a) shows opacification of a large inferior diaphragmatic vein without opacification of the gastric varices. Following partial embolization of the outflow veins, the gastric varices could still not be opacified from the gastrorenal shunt. As such, trans-splenic access was obtained (►Fig. 5b, black arrows) through which the gastric varices (►Fig. 5b, white arrow) were opacified with persistent outflow through the inferior diaphragmatic vein. Following complete embolization of the inferior diaphragmatic vein (►Fig. 5c) portions of the gastric varices were opacified from the gastrorenal shunt access site. Embolization with 3% Sotradecol (AngioDynamics, Inc., Queensbury, New York) embolic foam was performed in this portion of the gastric varices. Following initial Sotradecol foam (3 parts air:2 parts 3% STS:1 part ethiodized oil, Guerbet, Princeton, New Jersey) embolization (►Fig. 5d) injection from the trans-splenic access (►Fig. 5d, white arrows), the remaining gastric varices were opacified with drainage through the paraspinal venous plexus (►Fig. 5e). 2.5F Cantata (Cook Medical Inc, Bloomington, Indiana) microcatheter and 0.014 Synchro (Stryker, Kalamazoo, Michigan) microwire were used to access the primary outflow into the paraspinal venous plexus (►Fig. 5f). Following coil embolization in this location, the remaining gastric varices were embolized (►Fig. 5g). This patient's melena ceased immediately. He has since received a transplant liver.

### Anorectal Varices

Anorectal varices form from a feeding superior rectal vein and drained by either a middle or inferior rectal vein. Anorectal varices are reported to occur in up to 35% of pediatric patients with portal hypertension from both intrahepatic and prehepatic causes.<sup>44</sup> There is otherwise scant literature describing the epidemiology and prevalence of pediatric anorectal varices. In the authors' experience, the prevalence of anorectal varices is less than that reported in the literature. Based on available adult literature, there is an increased incidence of anorectal varices in patients with gastroesophageal varices. There is an additional increased incidence if a patients' gastroesophageal varices have previously hemorrhaged.<sup>45</sup> Although anorectal varices are more common in adults, they have a low clinically significant bleeding rate from 0.5% to 5%.<sup>46</sup> Anorectal variceal gastrointestinal hemorrhage can be difficult to detect and treat. There are currently no guidelines in the adult or pediatric literature regarding management of bleeding anorectal varices.

There are a variety of approaches to managing anorectal variceal hemorrhage, however, one of the initial steps include vasoactive agents (terlipressin or octreotide) in com-

ination with empiric antibiotic therapy.<sup>47</sup> In the event that endoscopic ligation or sclerosis cannot be performed, endovascular therapy can be an alternative option. Although TIPS and BRTO have been successful in the pediatric literature for gastroesophageal varices as described above, there are many successful modified techniques for managing bleeding anorectal varices in the adult literature including TIPS, double balloon-occlusion embotherapy (DBOE),<sup>48</sup> modified percutaneous transhepatic obliteration (MPTO),<sup>49</sup> BRTO,<sup>50</sup> balloon-occluded antegrade transvenous sclerotherapy (BATS), and balloon-occluded antegrade transvenous obliteration (BATO)<sup>51,52</sup> (►Fig. 6a,b). There are no published pediatric cases of the other modified techniques described in the adult literature, but given their technical success, these are potential options for managing bleeding anorectal varices. In addition to the variable techniques once selecting the varix in question, there have been multiple different adult cases published on the approach to reach the varix of interest including transhepatic, paraumbilical, and transileocolic access (through a pararectal approach) which are potential routes to select the varix in pediatric patients given their high incidence of extrahepatic portal vein occlusion.<sup>49,53,54</sup>



**Fig. 6a,b** A 17-year-old boy with idiopathic portal hypertension complicated by recurrent gastrointestinal bleeding secondary to rectal varices, presented with severe rectal bleeding following endoscopic biopsy. The patient's bleeding was complicated by concomitant portal vein thrombosis requiring TIPS creation (► Fig. 6a, black arrow). At the time of the TIPS creation, embolization of the rectal varices was performed. Through the TIPS, a 4F catheter (► Fig. 6a, dashed black arrow) was used to access a markedly enlarged inferior mesenteric vein (► Fig. 6a, white arrows). Through this access, a 5F Python (Applied Medical, Rancho Santa Margarita, California) balloon occlusion catheter (► Fig. 6b, white arrowhead) was used to occlude outflow while injecting Sotradecol embolic foam (3 parts air:2 parts 3% STS:1 part ethiodized oil, Guerbet, Princeton, New Jersey) which completely filled the rectal varices (► Fig. 6b, white arrows). Multiple 0.035 coils were deployed through the occlusion balloon catheter to stop outflow of embolic foam. The patient's rectal bleeding ceased immediately. He has not had rectal bleeding since.

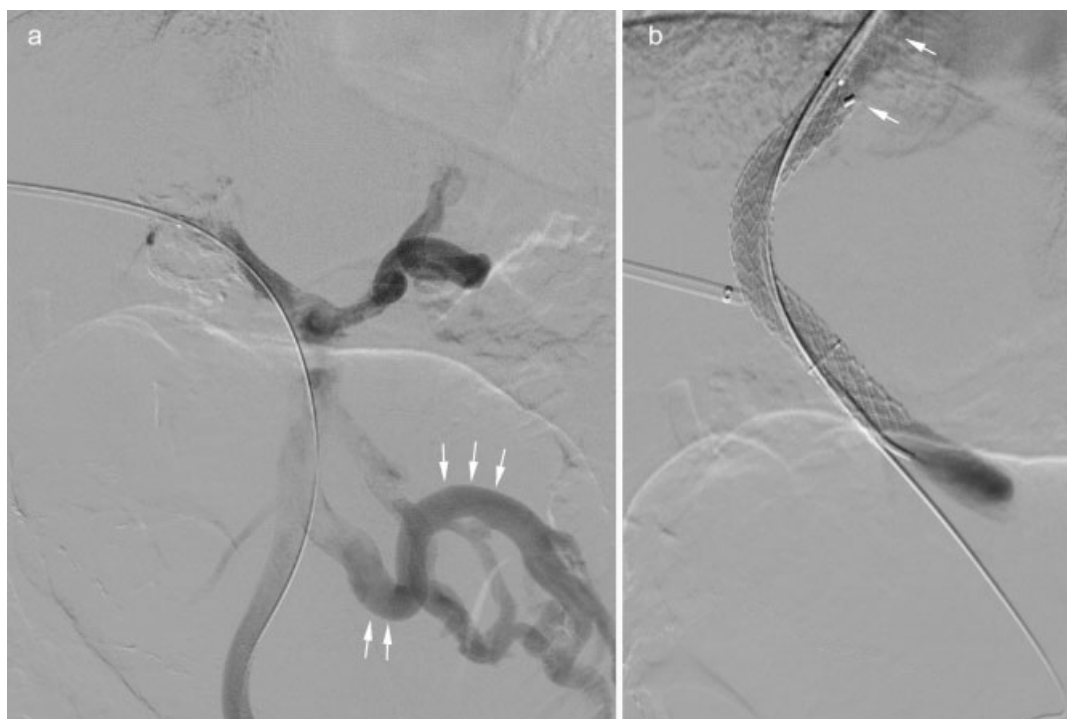
Embolic material is similar to that which can be used for gastric varices in BRTO as described above, and a single case report demonstrated success with a novel embolic and sclerosant mixture: 1 gm of Avitene (Bard, Tempe, Arizona), 4 mL of 3% Sotradecol (Angiodynamics, Latham, New York), and 1 cm<sup>3</sup> of shredded Gelfoam (Pfizer, New York), mixed in 40 mL of nonionic contrast medium (Omnipaque 350; GE Healthcare, Marlborough, MA) followed by an Amplatzer plug 2 in the inferior mesenteric vein.<sup>55</sup> Alternatively, embolization of reversed superior rectal venous tributaries, alone or in combination with TIPS, may be an effective temporizing measure. In all the adult cases, technical success was achieved without major complications; however, with any embolic deposition into the systemic venous circulation, there is a risk for nontarget embolization and pulmonary embolism. In the commonly used transhepatic approach, intraperitoneal hemorrhage has been reported to occur in up to 10.6% of adult patients.<sup>56</sup>

### Small Bowel and Roux Limb Varices

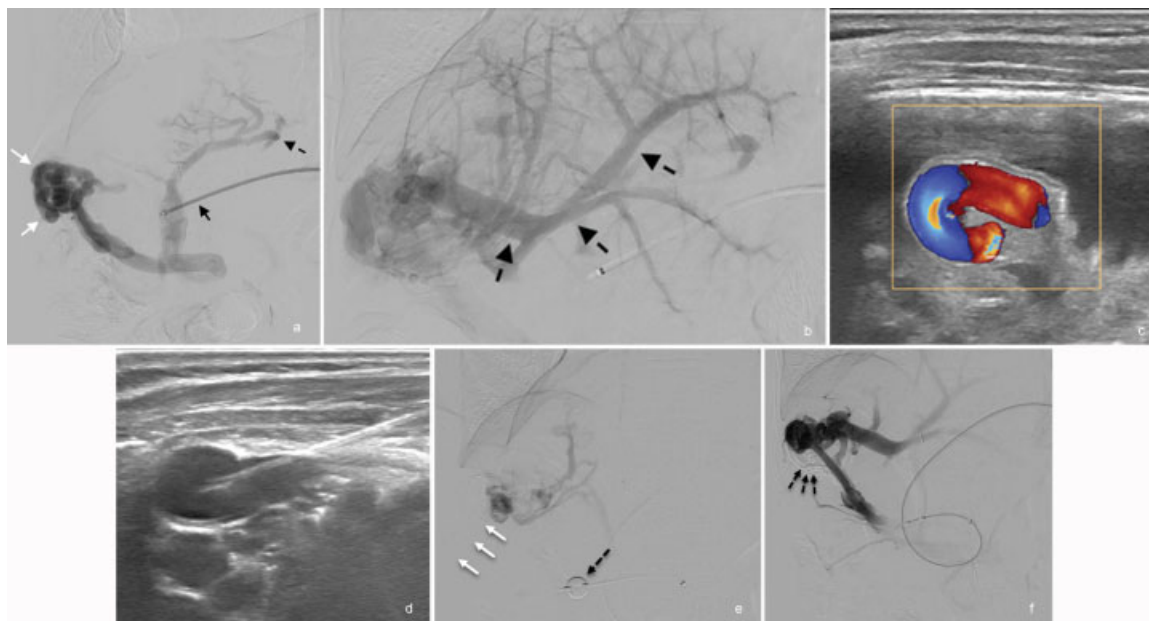
Pediatric small bowel varices are less common than other EcVs in the setting of portal hypertension and are difficult to diagnose; however, they can cause morbidity and mortality secondary to chronic gastrointestinal bleeding and severe anemia. It may be seen in a slightly higher frequency in children who have biliary atresia and have undergone a hepatopertoenterostomy who then proceed

with the development of portal hypertension.<sup>57</sup> When endoscopic evaluation fails to identify a source of upper or lower gastrointestinal bleeding, capsule endoscopy can aid in diagnosis and localization of ectopic small bowel varices for endovascular intervention.<sup>58</sup> Additional diagnostic imaging considerations include a conventional CT venous phase and percutaneous transhepatic portography.<sup>59</sup> Often, endoscopic management is not possible due to distal location when medical management fails.

Duodenal varices account for 17% to 33% of ectopic EcV,<sup>60-62</sup> and form as thin-walled retroperitoneal portoportal and/or portosystemic collaterals. The afferent feeding vessels include the pancreaticoduodenal, superior mesenteric, gastroduodenal, and pyloric veins while the draining vessels are typically the gonadal and capsular renal veins.<sup>63,64</sup> Two-thirds of duodenal varices originate from intrahepatic portal hypertension,<sup>65</sup> and although there is a relatively low prevalence of duodenal varices, there is a four-fold risk of bleeding when compared to esophageal varices.<sup>66</sup> There is no standardization of how to manage bleeding duodenal varices, and there is a paucity of literature regarding management of bleeding pediatric duodenal varices. In the adult literature, there are case reports and small case series demonstrating the technical success of endovascular management through TIPS (► Figs. 7a,b; 8a-c), BRTO, BATS, BATO, and improving portal venous patency.<sup>67,68</sup> The rebleed



**Fig. 7a,b** A 16-year-old boy with cystic fibrosis, end-stage liver disease, and long-standing portal hypertension with recurrent gastrointestinal bleeding and anemia. Capsule endoscopy demonstrated small bowel varices. Transhepatic access demonstrates a large ectatic superior mesenteric varix (► **Fig. 7a**, white arrows). TIPS was created with a 10 mm × 8 cm × 2 cm Viatorr (Gore, Flagstaff, Arizona) with subsequent cessation of gastrointestinal bleeding. Of note, the TIPS shunt was intentionally left short (► **Fig. 7b**, white arrows) so as not to interfere with future hepatic vein anastomosis in the setting of a liver transplant. Bleeding stopped following TIPS creation, and the patient received a liver transplant shortly thereafter.



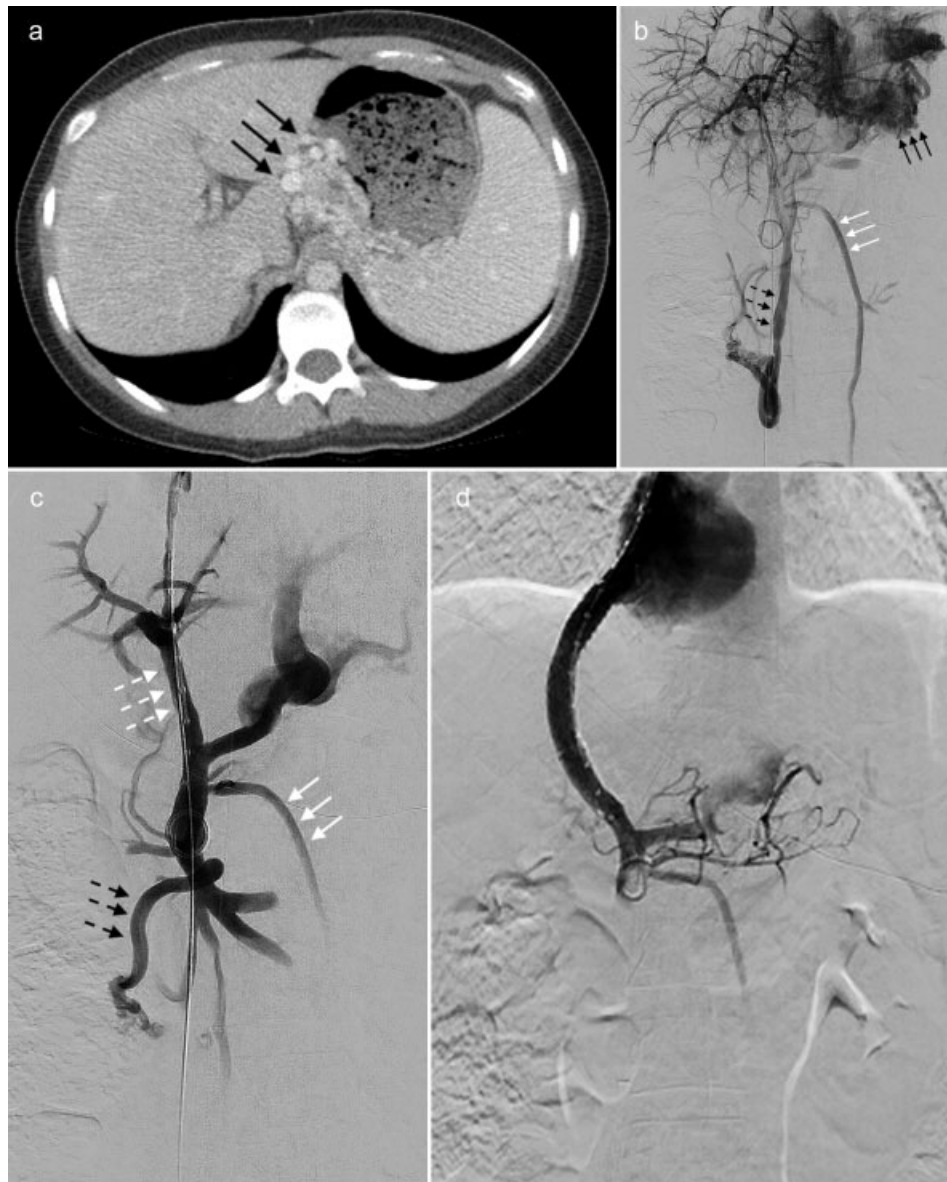
**Fig. 8a-f** A 14-year-old girl with cavernous transformation of the portal vein and severe gastrointestinal bleeding and anemia nonresponsive to transfusions many years after a left-lateral segment liver transplant for biliary atresia. Based on cross-sectional imaging, varices surrounding her roux-limb were suspected to be the source of bleeding. ► **Figure 8a** shows an injection from trans-splenic access (black arrow) filling large roux-limb varices (white arrows), as well as mesenteric-to-portal collaterals (black dashed arrows). ► **Figure 8b** shows a later phase following the same injection showing eventual opacification of the portal vein (dashed black arrows). Following multiple unsuccessful attempts to catheterize the roux-limb varices, ultrasound was used to identify the roux-limb varices (► **Fig. 8c**). Direct access was obtained into the varices (► **Fig. 8d**). ► **Figure 8e** shows contrast injection into the roux-limb varices through the percutaneous access (white arrows). Note the occlusion balloon (dashed black arrow), which was inflated to eliminate the risk of reflux of embolic foam into the native portal vein. Following injection of Sotradecol embolic foam (3 parts air:2 parts 3% STS:1-part ethiodized oil, Guerbet, Princeton, New Jersey) (► **Fig. 8f**, black dashed arrows), there is more rapid opacification of the intrahepatic portal vein with no enhancement of the roux-limb varices. Her bleeding ceased immediately.



rates of duodenal varices after TIPS is 21% to 37% and after transvenous obliteration is 13%.<sup>63</sup> A five-patient case series by Saad et al demonstrated a combination of TIPS and transvenous obliteration to be successful in preventing post-procedural duodenal variceal hemorrhage up to a mean of 22 months (range 6–50 months). Based on the nature of the thin-walled duodenal varices, the same authors recommend transvenous obliteration over coil embolization (newer softer coils preferred), and caution in avoiding the submucosal and extraluminal components of the varix to ensure adequate obliteration.<sup>68</sup> As with BRTO of the gastroesophageal varices, similar embolic agents have been used for obliterating duodenal varices including ethanolamine oleate, 3% sodium tetradecyl sulfate, 50% glucose, absolute etha-

nol,<sup>68</sup> and there are similar complications with BRTO as described above.

Jejunal varices can result in obscure bleeding and the prevalence is not well recorded in the literature. Only a single case of pediatric jejunal varix embolization has been reported, which was technically successful and in conjunction with trans-splenic portal vein recanalization and stenting after liver transplant.<sup>69</sup> A study of trans-splenic endovascular intervention (►Fig. 9a–f) demonstrated success in treating eight patients with Roux limb varices; however, intraperitoneal bleeding occurred in 27% of patients with a significant correlation between hemorrhage and intraprocedural anticoagulation.<sup>70</sup> The majority of published case reports and small case series of successful



**Fig. 9a–d** An 18-year-old female with portal sclerosis presented with upper gastrointestinal bleeding secondary to spontaneous portosystemic collaterals. Axial portal venous phase CT image (►Fig. 9a) demonstrated extensive gastroesophageal varices (black arrows). Early (►Fig. 9b) and late (►Fig. 9c) portal venogram prior to TIPS placement demonstrated a small caliber main portal vein (white dashed arrows), reversed flow within the inferior mesenteric vein (white arrows), an enlarged coronary vein supplying extensive gastric and gastroesophageal varices (black arrows), and duodenal varices draining to the inferior vena cava with the right gonadal vein (black dashed arrows). Post-TIPS portal venogram demonstrates decreased filling of the collaterals (►Fig. 9d).

management of bleeding jejunal varices in adults are in patients with prior liver transplant, extrahepatic portal venous obstruction, or hepatopancreatobiliary surgery (commonly choledojejunostomy) which suggests an altered venous collateralization pathway postprocedurally leading to varices at the jejunal anastomosis.<sup>59,71–75</sup> There are only three cases reported of adults patients with bleeding jejunal varices without portal hypertension or prior surgery,<sup>76</sup> and none in the pediatric literature.

In the single published pediatric patient with a bleeding jejunal varix, a Gelfoam slurry (Pfizer, New York), was used to achieve hemostasis,<sup>69</sup> while the adult literature expands on the treatment options with two main treatment options as direct jejunal variceal obliteration or portal vein decompression to improve hepatopetal flow, occasionally as sequential or combined procedures.<sup>73</sup> Successful embolization of small bowel varices in adults has been achieved with anhydrous ethanol, NBCA (NBCA:ethiodized oil 1:1–1:6), interlocking coils, and/or microcoils.<sup>59,71–73,75</sup>

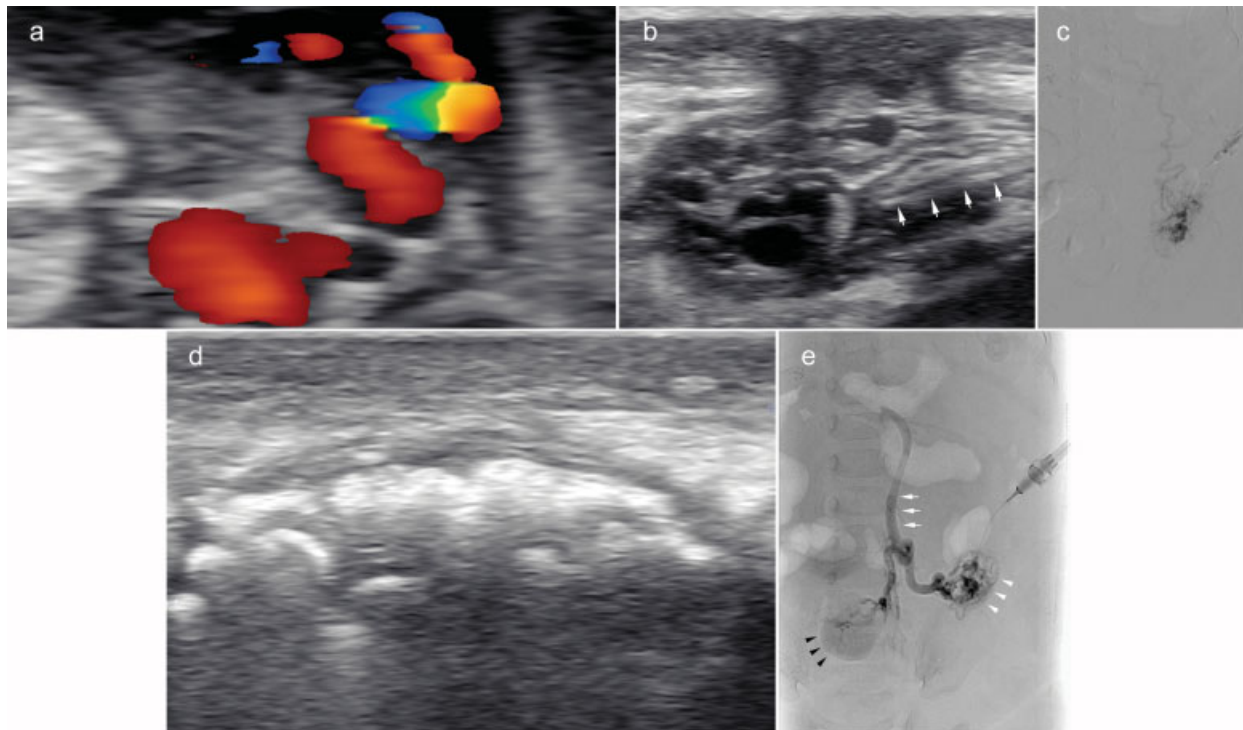
### Stomal Varices

Stomal varices occur at the surgically created mucocutaneous anastomosis secondary to portosystemic collateralization after colonic or small bowel enterostomies.<sup>63</sup> These types of varices have a higher association in patients with ulcerative colitis and primary sclerosing cholangitis.<sup>77</sup> Stomal varices hemorrhage in up to 27% of patients and occur on

average 2 years after stoma formation. Stomal variceal hemorrhage conveys a 3% to 4% mortality risk.<sup>78</sup> Typically, the feeding vessels of stomal varices are the colic or ileal veins, and the inferior epigastric is the dominant draining vessel.<sup>75</sup> Given their superficiality, stomal varices can be diagnosed with Doppler ultrasound and are commonly treated conservatively such as with direct pressure. For recurrent or life-threatening bleeding, more invasive treatment may be required. However, as with other EcV, there are no standard guidelines for management and the two mainstays of endovascular therapy are portal decompression (TIPS, improving hepatopetal flow) or direct embolization.<sup>79</sup> The pediatric literature is scarce in describing direct embolization techniques; however, small case series in the adult literature had successful obliteration with BRTO with NBCA:ethiodized oil (1:3–1:6), onyx, and coil embolization. Endovascular therapy (►Fig. 10a–e) has a high technical success rate with a low rebleeding rate in the multiple case studies in the adult literature.<sup>4,80–84</sup>

### Postprocedural Imaging

Postoperative imaging follow-up has not been well described in the pediatric setting for less common EcV discussed in this review. The adult literature describes post-BRTO imaging recommendations to confirm obliteration of the varices. Given the lack of data for other adult EcV follow-up, the recommendations below can be an alternative option for postprocedural



**Fig. 10a–e** A 7-month-old male child with biliary atresia and portal hypertension complicated by peristomal varices (►Fig. 10a) an intractable hemorrhage. Direct access was obtained into the peristomal varices using a 22-gauge angiocatheter (►Fig. 10b). Initial contrast injection (►Fig. 10c) showed drainage from the ileostomy peristomal varices through a small collateral vessel. This outflow vein thrombosed quickly, redistributing embolic foam (3 parts air:2 parts 3% STS:1-part ethiodized oil, Guerbet, Princeton, New Jersey) into the varices surrounding both the ileostomy and mucous fistula. ►Figure 8d shows ultrasound appearance of the embolic foam. ►Figure 10e shows distribution of embolic filling the varices around the ileostomy (white arrowheads), the varices near the mucous fistula (black arrowheads), as well as the inferior mesenteric vein. The patient had immediate cessation of bleeding and received a transplant a few weeks later.

imaging for all types of treated EcV until tailored recommendations are further defined. Prior to discharge or within the first week, it is recommended to obtain contrast-enhanced cross-sectional imaging and/or endoscopic ultrasound imaging of the varices to ensure obliteration and assess for complications such as portal thrombus, retroperitoneal bleeding, and infection.<sup>85</sup> Long-term follow-up is recommended with contrast-enhanced computed tomography (CECT) or contrast-enhanced magnetic resonance imaging (CEMRI) at 1, 3, 6, and 12 months, then every 6 months or annually.<sup>86,87</sup> When ethiodized oil is used to aid in visualization of the foam forms of the sclerosant during angiography, follow-up with CEMRI may be considered secondary to ethiodized oil's hyperdense appearance on CECT.<sup>88</sup> Endoscopic ultrasound may provide additional diagnostic value with the ability to characterize thrombosed or partially thrombosed gastroesophageal varices, predict risk of specific variceal bleeding, and identify collateral vasculature information for endoscopic treatment. However, current limitations include cost and availability of equipment and expertise at pediatric hospitals.<sup>89,90</sup>

## Conclusion

Pediatric portal hypertension can lead to portosystemic collateralization in the form of gastroesophageal, isolated gastric, anorectal, small bowel, roux-limb, and stomal varices. These varices can result in morbidity and mortality secondary to acute and/or chronic bleeding that can be life threatening. Although TIPS for bleeding gastroesophageal varices has been the most well-studied endovascular or percutaneous therapy in the setting of EcV, little literature is otherwise available to guide clinicians caring for patients with bleeding EcV in other locations. To date, the majority of the literature is derived from case reports and small case series to describe the management of these bleeding ectopic varices, and the literature demonstrates a lack of standardization in management. This review demonstrates that percutaneous endovascular therapeutic options can be safe and efficacious options to embolize ectopic varices in appropriately selected pediatric patients when medical and endoscopic therapy fail or are precluded.

### Conflict of Interest

None.

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