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Management and classification of type II congenital portosystemic shunts☆

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Abstract

Background: Congenital portosystemic shunts (PSS) with preserved intrahepatic portal flow (type II) present with a range of clinical signs. The indications for and benefits of repair of PSS remain incompletely understood. A more comprehensive classification may also benefit comparative analyses from different institutions.

Methods: All children treated at our institution for type II congenital PSS from 1999 through 2009 were reviewed for presentation, treatment, and outcome.

Results: Ten children (7 boys) with type II PSS were identified at a median age of 5.5 years. Hyperammonemia with varying degrees of neurocognitive dysfunction occurred in 80%. The shunt arose from a branch of the portal vein (type IIa; n = 2), from the main portal vein (type IIb; n = 7), or from a splenic or mesenteric vein (type IIc; n = 1). Management included operative ligation (n = 6), endovascular occlusion (n = 3), or a combined approach (n = 1). Shunt occlusion was successful in all cases. Serum ammonia decreased from 130 ± 115 μmol/L preoperatively to 31 ± 15 μmol/L postoperatively ($P = .03$). Additional benefits included resolution of neurocognitive dysfunction (n = 3), liver nodules (n = 1), and vaginal bleeding (n = 1).

Conclusion: Correction of type II PSS relieves a wide array of symptoms. Surgery is indicated for patients with clinically significant shunting. A refined classification system will permit future comparison of patients with similar physiology.

Congenital absence of the portal vein with an end-to-side portocaval shunt was first described by John Abernethy in 1793 [1]. Many variants of congenital portosystemic shunts (PSS) have been subsequently described, and in 1994, Morgan and Superina [2] introduced a classification system based on whether the portal vein, often hypoplastic, was present and whether the liver was perfused with blood from the mesenteric venous system. In a type I shunt, there is a complete end-to-side portocaval fistula with no discernable portal flow to the liver. Type II shunts occur as a side-to-side portocaval fistula or as any number of other PSS including gastrorenal, splenorenal, and portorenal shunts. The key feature of type II shunts is preservation of at least some hepatic portal flow. In 1997, Howard and Davenport [3] applied the Abernethy eponym to portocaval shunts and, again, recognized the type I and type II variants. The many

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anatomic variations of these shunts have been recently detailed [4].

Congenital absence of the portal vein (type I Abernethy malformation) occurs more frequently in girls and is associated with other congenital anomalies [3,4]. Liver transplantation, which offers the only option for cure, is reserved for patients with refractory symptoms despite medical management. Type II PSS, on the other hand, have considerably more variability for anatomy, clinical features, and treatment options. Hyperammonemia, which is due to shunting of blood away from the portal circulation, has been the most commonly reported problem [5,6]. Subtle symptoms of encephalopathy may not manifest until adulthood [5,7]. Regenerative liver lesions and pulmonary hypertension have also been associated with type II shunts [3,4,7-9]. Operative ligation and/or endovascular coiling of these fistulas have been described in case reports and small series, but the risks vs benefits of intervention remain to be determined [6,8]. We report the benefits and potential pitfalls of operative and endovascular intervention in our experience in managing 10 patients with type II congenital PSS. A refinement of the classification for type II shunts is proposed to facilitate clinically relevant discussion of the anatomy, physiology, and management.

1. Methods

All patients treated for type II congenital PSS at Children's Memorial Hospital (Chicago, IL) from 1998 through 2009 were retrospectively reviewed. Inpatient and outpatient medical records were queried to determine demographic information, presenting symptoms, associated medical problems, operative course, and clinical outcome. The study was approved by the hospital's institutional review board. Continuous variables are reported as mean ± SD as well as median values. Comparison of continuous variables from before and after surgery was performed using paired t test, and $P < .05$ was considered significant.

Shunt anatomy was classified according to the origin of the fistula, regardless of whether intrahepatic or extrahepatic (Table 1). A shunt arising from a branch of the portal vein was classified as type IIa. This includes the patent ductus venosus (PDV), which connects the left portal vein to the left hepatic vein near its entry into the inferior vena cava (IVC). Shunts arising from the main portal vein or its bifurcation were classified as type IIb, whereas those arising from the mesenteric, gastric, or splenic veins were classified as type IIc.

Table 1 Classification of congenital PSS

Type	Description
I	No intrahepatic portal flow (CAPV or type I Abernethy malformation)
II	Partial shunt with preserved hepatic portal flow (type II Abernethy malformation)
IIa	Arising from left or right portal vein (includes PDV)
IIb	Arising from main portal vein (including its bifurcation or splenomesenteric confluence)
IIc	Arising from the mesenteric, gastric, or splenic veins

CAPV indicates congenital absence of the portal vein.

1.1. Operative technique

The endovascular techniques used at our institution for closure of appropriate PSS have been previously described [9]. The operative algorithm for patients requiring laparotomy includes the following steps. At the time of initial exploration, a catheter is introduced into the portal system via a small jejunal branch to transduce mesenteric venous pressure. The shunt is then isolated and temporarily occluded. The change in venous pressure before and after shunt occlusion is noted. In addition, a Doppler flow probe is used whenever possible to measure portal vein blood flow before and after occlusion. Finally, an intraoperative portal venogram is performed to confirm the filling of intrahepatic portal venous branches. If the rise in portal pressure is deemed acceptable (<25 mm Hg absolute pressure), the appearance of the bowel is not worrisome, and there is an accompanying increase in portal venous flow, then the abnormal venous fistula may be permanently occluded. Otherwise, a doubly looped vessel loop secured by a clip is left in place to partially or completely occlude the shunt, and the patient is transferred to the intensive care unit with a temporary abdominal closure and the venous catheter for portal pressure transduction left in situ. The patient is placed on heparin drip to avoid thrombosis in the low-flow mesenteric venous system. During the ensuing days, portal pressure is monitored continuously, while the intrahepatic portal system is allowed to expand and accommodate the increase in pressure. Transabdominal Doppler ultrasonography can be performed as needed to monitor portal flow. After 3 to 5 days, a portal venogram is performed via the existing catheter to confirm expansion of the intrahepatic portal system. If the intrahepatic portal veins have dilated sufficiently and portal pressures are tolerable, the shunt is permanently ligated. Otherwise, the fistula is left partially banded with a vessel loop secured to achieve maximal shunt constriction without increasing portal pressure beyond 20 mm Hg.

2. Results

Ten children (7 boys) with type II congenital PSS were identified (Table 2). The median age at presentation was 5.5 years (range, 0.5-16 years). Hyperammonemia was the most frequent problem (cases 1-4, 7, 8, and 10), and the mean preoperative ammonia was 130 ± 115 μmol/L

Table 2 Clinical features, shunt classification, management, and outcome of children with congenital PSS

Case no.	Age (y)/ sex	Presenting symptoms	Ammonia (μmol/L) Pre | Post	Fistula classification and anatomy	Procedure(s)	Outcome	Follow-up duration
1	1/M	Jaundice and hyperammonemia	102 | 48	Type IIb (portocaval)	Operative (initial ligation)	Bilirubin and ammonia normalized	10 mo
2	10/ M	Coagulopathy with hematuria and liver lesions	86 | 9	Type IIb (portocaval)	Operative (initial ligation)	Liver lesions, coagulopathy, and ammonia resolved	36 mo
3	0.5/ M	Coagulopathy and hyperammonemia	84 | 26	Type IIb (portocaval)	Operative (staged ligation)	Coagulopathy and ammonia normalized	19 mo
4	8/M	Neurocognitive dysfunction and seizures	145 | 33	Type IIb (portocaval)	Operative (initial ligation)	Seizures resolved and subjective improvement in neurocognitive function	20 mo
5	7/F	Vaginal bleeding	38 |	Type IIc (mesoiliac)	Hybrid (ligation followed by embolizations)	Vaginal bleeding resolved	10 mo
6	8/F	Neurocognitive dysfunction and seizures	| 27	Type IIb (PV to RA)	Endovascular (closure device)	Subjective improvement in neurocognitive function	14 mo
7	16/ F	Neurocognitive dysfunction and regenerative liver nodules	110 | 61	Type IIa (PDV and small additional shunts)	Endovascular (multiple embolizations)	Small residual shunt and objective improvement in neurocognitive function	36 mo
8	3/M	Protein-losing gastropathy	73 | 13	Type IIb (portocaval)	Operative (staged ligation)	Ammonia normalized	10 mo
9	4/M	Neurocognitive dysfunction		Type IIa (PDV)	Endovascular (concentric stents)	Objective improvement in neurocognitive function	24 mo
10	2.5/ M	Hyperammonemia	400 | 45	Type IIb (portocaval)	Operative (partial banding)	Ammonia normalized	5 mo

M indicates male; F, female; PV, portal vein; RA, right atrium.

(median, 94 μmol/L; reference range, 11-35 μmol/L). Treatment of hyperammonemia with severe restriction of dietary protein consumption, lactulose, or gut antibiotic administration had been attempted in most cases. Four patients had documented neurocognitive dysfunction (cases 4, 6, 7, and 9). Other associated symptoms included seizure activity (cases 4 and 6), coagulopathy (cases 2 and 3), failure to thrive (case 3), and regenerative liver nodules (cases 2 and 7). One girl with a mesoiliac fistula presented with vaginal bleeding (case 5). The fistula arose from a branch of the portal vein (type IIa) in 2 patients (cases 7 and 9), from the main portal vein or its bifurcation (type IIb) in 7 patients (cases 1-4, 6, 8, and 10), and from a mesenteric vein (type IIc) in 1 patient (case 5).

Preoperative evaluation included ultrasound in 7 patients, computed tomographic (CT) angiogram in 7 patients, and magnetic resonance in 1 patient. Preoperative venogram was performed in 6 patients (cases 1, 2, 4, 6, 9, and 10). Measurement of portal pressure with trial shunt occlusion (either endovascular or intraoperative) was performed in all 9 patients with portocaval fistulas. Portal pressure measurement was not indicated in the patient (case 5) with a mesoiliac fistula. Of 9 patients, 5 had an acute rise in portal pressure (>20 mm Hg) with temporary shunt occlusion (cases 3 and 7-10) and, therefore, required staged procedures (2 endovascular and 3 open). Case 4 had an acute rise in portal pressure, which stabilized after a short period of observation in the operating room, and he tolerated definitive shunt ligation at the initial operation. Cases 1 and 2 tolerated immediate shunt ligation, and case 6 tolerated shunt embolization with no rise in portal pressure.

Final management was therefore operative (n = 6), endovascular (n = 3), or combined (n = 1). Among the 3 patients who underwent definitive endovascular procedures, 1 tolerated immediate placement of an occlusion device in a long narrow shunt from the portal vein to the right atrium (case 6). The other 2 patients had PDV (cases 7 and 9). One required staged embolizations because of elevated portal pressure. She also needed additional endovascular procedures over the subsequent 2 years to obtain occlusion of the ductus venosus and to manage additional collateral shunts, which developed. Sequential, concentric endovascular stents were used to gradually close the PDV in the other patient. A combined approach was used in a girl with vaginal bleeding that persisted after open ligation of a dominant fistula between the IMV and an iliac branch (case 5). Vaginal bleeding stopped after additional mesoiliac fistulas were coiled.

Perioperative complications occurred in 2 patients. Case 4 developed IVC stenosis, which was successfully managed

with placement of an expandable covered stent in the IVC as well as a portal vein thrombus, which resolved with systemic anticoagulation. Case 8 had a large portocaval fistula and a threadlike extrahepatic and intrahepatic portal venous system. He underwent staged shunt closure over multiple operations with continuous portal pressure measurement but developed portal vein stasis and partial thrombotic occlusion of the superior mesenteric vein. Catheter-directed thrombolysis with tissue plasminogen activator in addition to systemic heparinization was successful at restoring portal flow. However, the boy developed a limited motor deficit from an intracranial bleed, requiring cessation of all antithrombotic measures. The focal motor deficit improved significantly over the course of his hospitalization. His intrahepatic portal system progressively dilated over the course of several weeks. He is doing well with minimal residual neurologic deficit 10 months later.

Shunt occlusion with symptom resolution was ultimately obtained in all 10 patients. Serum ammonia decreased from 130 ± 115 μmol/L preoperatively to 27 ± 15 μmol/L (median, 26) postoperatively ($P = .03$). Dietary restrictions to reduce serum ammonia levels were lifted in all patients. The objective improvement in neurocognitive function in the 2 patients with PDV who underwent endovascular shunt closure has previously been reported (cases 7 and 9) [9]. Learning difficulties improved subjectively in 2 patients (cases 4 and 6), and seizure activity resolved in 1 patient (case 4). The liver lesion regressed in the patient with evidence of regenerative nodules preoperatively (case 2) (Fig. 1). Vaginal bleeding ceased in case 5. Follow-up imaging showed improved intrahepatic flow in all patients. Median postoperative follow-up was for 17 months (5-36 months).

3. Discussion

Review of our experience in managing children with type II congenital PSS illustrates the benefits of occluding the abnormal communication. Shunt anatomy is highly variable with resultant differences in clinical presentation and optimal operative approach. Successful shunt closure can alleviate symptoms of hyperammonemia and related neurologic symptoms, result in resolution of regenerative liver nodules, reverse growth impairment, and improve coagulopathy in affected patients.

It is increasingly recognized that the difference between type I and type II shunts is really more of a continuum than an absolute distinction [4]. Our experience would indicate that intrahepatic portal flow can be restored in children with all but the most profoundly hypoplastic veins. We have developed a management strategy for children with large shunts arising from the main portal vein, its bifurcation, or at the confluence of the splenic and superior mesenteric veins, as occurred in 6 patients in this study. Many of these patients have extremely hypoplastic portal veins distal to the shunt that are sometimes difficult to visualize with conventional CT angiography. In these patients, direct catheterization of the shunt by interventional radiology is essential to study the anatomy of the shunt and, whenever possible, conduct a trial occlusion of the shunt with pressure measurements in the proximal mesenteric veins to assess the physiologic consequences on the mesenteric venous pressure of sudden cessation of flow through the shunt. This knowledge is useful in planning operative strategy and aids in discussing the treatment pathway with the family of the child. It appears from our experience that there is a direct correlation between the severity of the hypoplasia of the portal vein and the position of the fistula along the mesenteric venous system: more proximally located shunts produce more severe hypoplasia of the portal vein and its intrahepatic branches and a more complete diversion of mesenteric venous blood into the systemic circulation.

The operative algorithm described herein has allowed for successful closure of portocaval shunts in children with extremely diminutive intrahepatic portal veins. Temporary shunt occlusion is very useful in distinguishing between patients who can tolerate immediate shunt ligation and those who develop severe portal hypertension and require a staged approach. We and others have had success using partial shunt banding to allow gradual expansion of the intrahepatic portal system while avoiding catastrophic portal hypertension. After the intrahepatic portal veins have dilated sufficiently and the portal pressure normalized, the residual shunt may close spontaneously, as in one of our cases (case

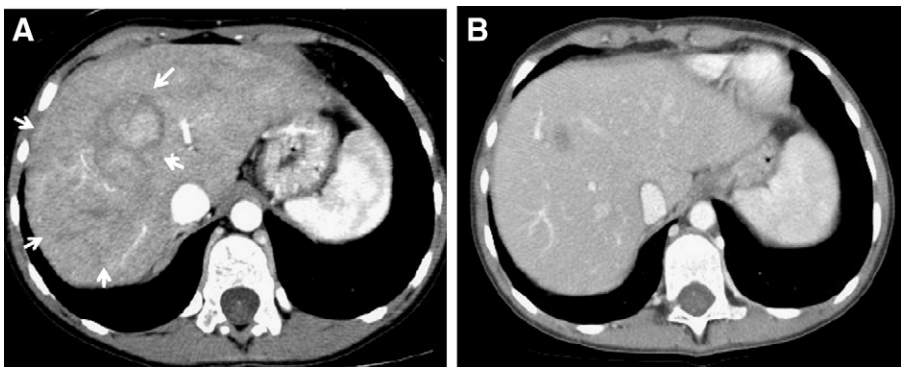

Fig. 1 Regenerative liver lesion (arrowheads) (A) in a child with a portosystemic shunt, which resolved after shunt closure (B).

10). Otherwise, a second operation or interventional procedure can be performed after several months to complete the shunt closure. However, we have found that, in many cases, temporary occlusion of the shunt for several days in a closely monitored environment allows sufficient intrahepatic portal expansion to permit definitive closure during a single hospitalization (Fig. 2). Further data are needed to determine the optimal operative approach and timing for patients with this very challenging problem.

Ongoing efforts to optimize the management of children with congenital PSS require standardized terminology. The simple distinction between type I and type II shunts remains critical for distinguishing patients who may benefit from nontransplant intervention [2]. Subsequent efforts to subclassify type II shunts have been fraught with confusion, redundancy, and a lack of clinical relevance. Distinction between intrahepatic and extrahepatic shunts is an overly subjective measure, as in cases of predominantly extrahepatic shunts that traverse a section of caudate lobe. Patent ductus venosus refers specifically to a persistent opening of the embryonic connection between the left portal vein and the left hepatic vein, but this term is frequently misused in reference to all types of intrahepatic shunts. Finally, anatomic classification of intrahepatic shunts using the separate classification of Park et al [10] generates confusion and unnecessary anatomic distinction without clinical applicability. In contrast, the classification of type II PSS based on portal anatomy proposed herein is simple and should be readily understandable. This classification provides insight into the expected physiologic consequences of the shunt and may have implications for management strategies.

3.1. Type IIa PSS (arising from a portal branch)

Shunts arising from the left or right portal vein including PDV are classified as type IIa. Although predominantly intrahepatic, the extent of liver parenchymal involvement is inconsequential. As expected, shunt fraction may be less than in those arising from the main portal vein [11]. Nonetheless, shunt fraction varies considerably, and hyperammonemia is still a problem in many children. Type IIa shunts are unique in that spontaneous closure has been reported in at least 13 young children [4,12,13]. When persistent, endovascular closure using coils, plugs, and/or stents has proven successful for many children with PDV [9,14,15] and other isolated type IIa shunts [16,17]. Liver mobilization typically allows for ligation of type IIa shunts without liver resection when coiling is not possible [18-20].

At least 56 type IIa shunts are reported in the literature, including 36 PDV [3,4,6,8,9,14,15,19-23]. Seventy-four percent of patients were boys, and the median age at diagnosis was 2.4 years. Among 29 patients who underwent

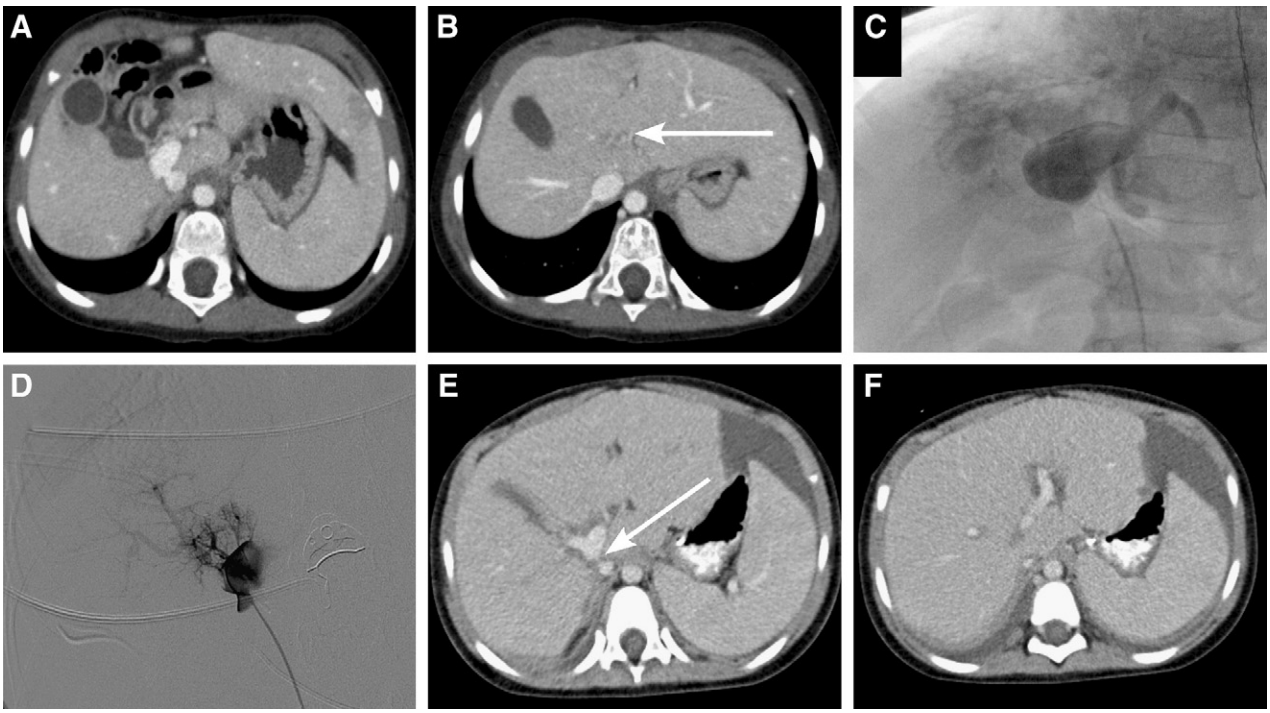

Fig. 2 A, A type IIb portocaval fistula with a profoundly hypoplastic intrahepatic portal system is seen on preoperative CT scan. B, The intrahepatic portal branches are not visualized in their expected location (arrow). C, Intraoperative venogram demonstrates a paucity of distinct intrahepatic portal branches. D, Distinct portal branches begin appearing 2 days after shunt occlusion. Computer tomographic scan 2 weeks after surgery demonstrates the absence of a fistula in its former location (arrow) (E) and improved opacification of the intrahepatic portal branches as well as a decrease in the caliber of the IVC (F). Some ascites, as seen here, is not uncommon in the early postoperative period when there is a transient rise in portal pressure.

operative (n = 15) or endovascular (n = 14) closure, 13 of 13 with encephalopathy or hyperammonemia improved after surgery, and 10 of 12 with cardiopulmonary symptoms improved after surgery. Further efforts are needed to determine at which age spontaneous closure of type IIa shunts becomes unlikely and which patients benefit from operative vs endovascular closure.

3.2. Type IIb PSS (arising from main portal vein, its bifurcation, or the splenomesenteric confluence)

Shunts arising from a position between the bifurcation of the portal vein and the splenomesenteric confluence can be classified as type IIb. These include main portocaval, portoatrial, and portorenal shunts. Although predominantly extrahepatic, these shunts may traverse a short segment of liver parenchyma. These abnormalities may be limited to a single communication from the main portal vein to the IVC. However, they may also compose a large, complex, and hard-to-delineate venous confluence of mesenteric and systemic veins encompassing the splenic, superior mesenteric, portal, and renal veins and the IVC. Hyperammonemia and other metabolic consequences are frequent. Type IIb shunts are typically short and wide in diameter, making successful endovascular management unlikely. Nonetheless, successful endovascular closure has been described in rare cases [6,16,24], including our patient with a long fistula draining into the right atrium. These patients usually require staged operative closure using the above-described technique. The degree of difficulty in managing these patients is directly related to the extent of portal vein hypoplasia and the location of the shunt. More proximal shunts (at or near the splenomesenteric confluence) often result in a long segment of hypoplastic extrahepatic portal vein and greater difficulty in restoring intrahepatic portal flow without causing excessive portal hypertension or stasis.

Including our patients, at least 26 cases of type IIb shunts have been reported [3,4,6,7,16,25-31]. Median age at diagnosis was 2.8 years, and 16 (62%) were boys. Shunt anatomy was portocaval in 10, portorenal in 5, and portoatrial in 1. Correction was attempted in 15 (3 endovascular, 10 open, and 1 laparoscopic) and was successful in 13. One child was intolerant of shunt banding, and another developed a new PSS after initial shunt ligation. All but 1 patient who underwent surgery had hyperammonemia with or without encephalopathy, and this resolved in all patients with successful shunt closure. In addition, liver lesions regressed in 2 patients, and cardiopulmonary symptoms improved in 2 patients after successful surgery.

3.3. Type IIc PSS (shunt arising from mesenteric, gastric, or splenic veins)

A wide variety of peripheral PSS can be grouped together as type IIc [8,28,29,32-35]. These shunts arise from the mesenteric, gastric, or splenic veins and empty into the renal vein, azygos vein, iliac veins, or their branches. The most frequent anatomic variants in children are gastrorenal (4 reported cases) and splenorenal (4 reported cases). Although more peripheral, encephalopathy or subclinical hyperammonemia nonetheless occurred in 10 of 14 reported cases. Cardiopulmonary symptoms occurred in 6 cases, and isolated cases of liver lesions and failure to thrive were reported. A shunt between the IMV and a branch of the iliac vein caused recurrent vaginal bleeding in 1 patient from our series. Some degree of portal vein hypoplasia can occur but usually to a lesser extent than in type IIb shunts. Therefore, immediate ligation or occlusion of type IIc shunts can typically be performed without causing severe portal hypertension or other adverse sequelae. Furthermore, these shunts may be approachable by minimally invasive techniques. Among the reported cases, 3 endovascular, 3 laparoscopic, 3 open, and 1 combined approaches were all successful.

In conclusion, symptoms caused by type II PSS are generally reversible with shunt ligation or embolization, and an aggressive approach aimed at elimination of the abnormal portosystemic communication should be taken. The approach for occlusion of the shunt must be based on the individual characteristics of the patient's shunt anatomy and position. Shunt closure is ultimately possible and tolerated even in patients with profoundly hypoplastic intrahepatic and extrahepatic portal veins using a staged procedure described herein. A refined classification based on portal anatomy is proposed, which will allow future studies to make meaningful comparisons between patients with similar anatomy and physiology.

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