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Review Article

Congenital Portosystemic Shunts: A Review

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Abstract

Congenital portosystemic shunts (CPSS) are rare vascular anomalies characterized by abnormal communication between the portal and systemic venous systems, resulting in partial or complete diversion of portal blood away from the liver. These shunts can give rise to a broad spectrum of clinical manifestations, including hyperammonemia (with or without encephalopathy), hepatopulmonary syndrome, and portopulmonary hypertension. Notably, these complications often occur in the absence of portal hypertension. Advances in diagnostic imaging, particularly Doppler ultrasound, computed tomographic angiography, and magnetic resonance imaging, have enhanced the early detection and classification of CPSS. Treatment approaches vary depending on shunt type and clinical severity and may include interventional closure via embolization or surgical ligation. Most persistent or symptomatic shunts require immediate intervention. Recent studies have also identified potential genetic and embryological mechanisms contributing to CPSS development, offering new insights into their pathogenesis. This review aims to summarize current knowledge on the epidemiology, pathophysiology, clinical presentation, diagnostic evaluation, and management of CPSS, and to highlight their consideration in patients with hepatic encephalopathy or unexplained liver disease.

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Introduction

Congenital portosystemic shunts (CPSS) are rare vascular anomalies resulting from aberrant fetal vascular development, leading to abnormal communications between the portal and systemic venous systems.[1] These connections divert portal blood away from the liver, resulting in reduced hepatic perfusion, impaired liver development, and altered metabolic processing.[2] CPSS are broadly classified into intrahepatic portosystemic shunts (IHPSS) and extrahepatic portosystemic shunts (EHPSS), the latter historically referred to as Abernethy malformations.

The estimated incidence of CPSS ranges from 1 in 30,000 to 1 in 50,000 per live births, although the true prevalence remains uncertain due to underdiagnosis and variable clinical presentation.[3] CPSS are diagnosed by Doppler ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI), either as part of the evaluation for congenital heart disease or syndromic conditions, or following the onset of clinical symptoms characteristic of CPSS.[4,5] This review aims to summarize current knowledge on the epidemiology, pathophysiology, clinical presentation, diagnostic evaluation, and management of CPSS, and to highlight their consideration in patients with hepatic encephalopathy but without cirrhosis.

Pathogenesis

Genetics

Gene expression is believed to play a significant role in the development of CPSS. Studies in dogs with IHPSS have identified impaired signaling in the aryl hydrocarbon receptor (AHR) pathway, a key regulator of ductus venosus closure. A breed-specific 6.3 kb LINE-1 insertion in intron 2 of the AHR gene is associated with reduced AHR expression and downstream alterations, particularly decreased HSP90AA1, which may impair nuclear translocation and stability of AHR and HIF1A. These molecular changes likely contribute to delayed or failed ductus venosus closure, resulting in persistent ductus venosus (Fig. 1C), supporting a polygenic, possibly digenic, inheritance model.[6]

Van Steenbeek *et al*. reported that IHPSS predominantly occurs in large-breed dogs, while EHPSS are more common in small-breed dogs, suggesting distinct genetic mechanisms underlying each type. In a comparative transcriptomic study, Van Steenbeek and colleagues found that although both shunt types share similar clinical consequences, they exhibit distinct hepatic gene expression profiles. IHPSS were characterized by increased expression of vascular cell adhesion molecule 1 (VCAM1) and the cell-cycle regulator WEE1, along with decreased expression of acyl-CoA–binding protein, cysteine conjugate–β-lyase 1, hepcidin, and palladin. In contrast, EHPSS showed reduced VCAM1 expression and decreased cysteine conjugate–β-lyase 1 levels, but not the WEE1 upregulation seen in IHPSS. These differences suggest that IHPSS and EHPSS arise through separate developmental pathways, possibly involving angiogenesis-related mechanisms in IHPSS and aberrant vitelline vein remodeling in EHPSS, highlighting VCAM1 and WEE1 as potential candidate genes for further study.[6]

Keywords: Congenital portosystemic shunts; Intrahepatic portosystemic shunts; Extrahepatic portosystemic shunts; Liver tumor; Endovascular approach; Liver transplantation.

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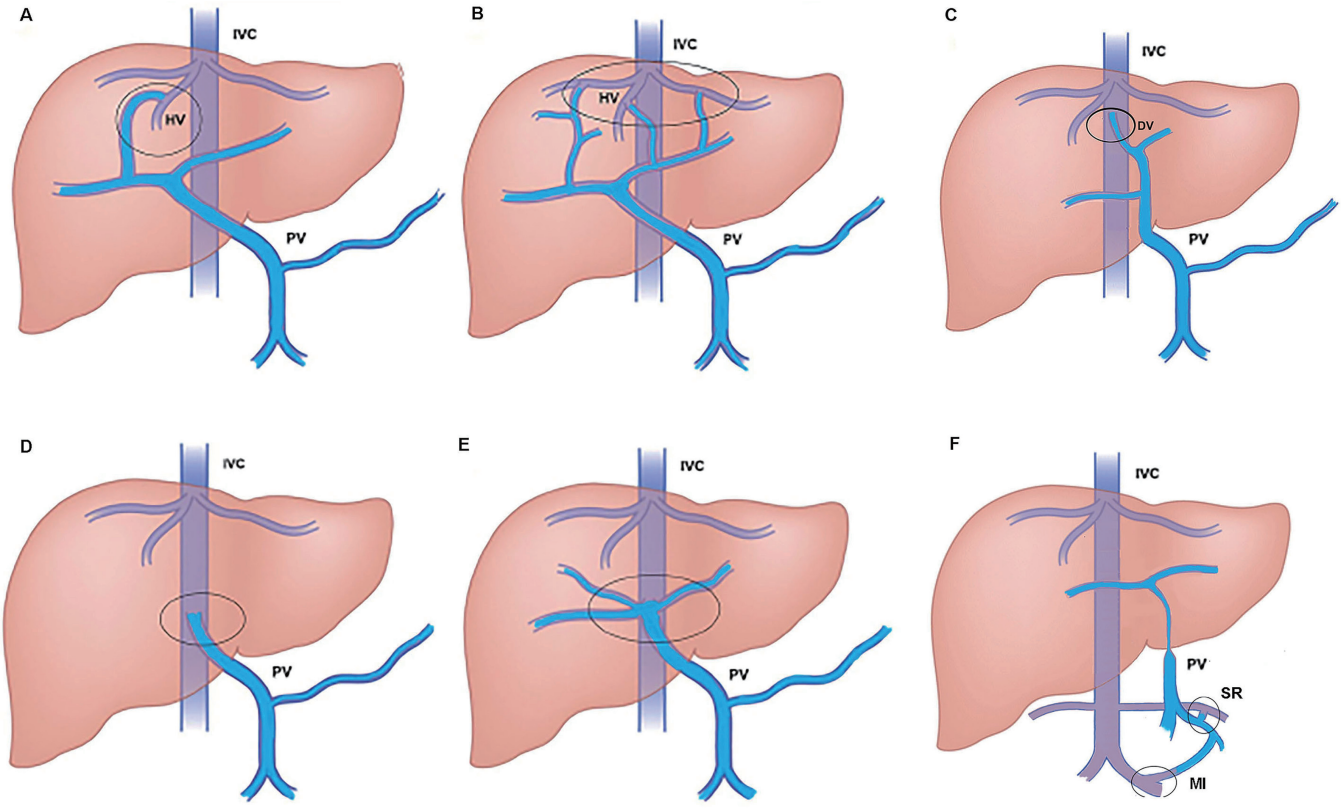

Fig. 1. Schematic diagrams of congenital portosystemic shunts illustrating the main anatomical subtypes. Panel A shows a single intrahepatic shunt connecting a portal vein branch to a hepatic vein, while Panel B demonstrates multiple intrahepatic shunts with diffuse communication between intrahepatic portal and systemic veins. Panel C depicts a patent ductus venosus shunting blood to the vena cava. Panel D shows an extrahepatic end-to-side shunt forming a direct connection between the main portal vein and the inferior vena cava, effectively bypassing hepatic circulation. Panel E illustrates an extrahepatic side-to-side shunt with preserved portal branching and a parallel conduit between the portal and systemic venous systems. Panel F shows examples of extrahepatic upstream shunts, including splenorenal and mesenteric iliac shunts formed secondary to stenosis of the portal vein. Light blue vessels represent portal blood flow. Dark blue vessels represent systemic venous flow. PV, portal vein; IVC, inferior vena cava; HV, hepatic vein; SR, splenorenal shunt; MI, mesenteric iliac shunt. Black circles highlight the vessels involved in shunts. (Adapted from Bahadori *et al*.[3])

The association of CPSS with other human congenital anomalies, such as heterotaxy, congenital heart disease, and chromosomal microdeletions, further supports a genetic component to its pathogenesis, suggesting a shared developmental and genetic basis for abnormal portal–systemic venous remodeling. These patterns indicate that the condition likely arises from disruptions in embryonic development rather than isolated events occurring after birth.[7–9]

Embryology

CPSS are believed to originate from disruptions in the normal embryological development of the abdominal venous system, which begins in the fourth week to the sixth week of gestation. During this period, the primordial liver develops and establishes connections with three major venous networks: the umbilical veins, the cardinal veins, and the vitelline veins. The cardinal veins contribute to the systemic venous system, while the vitelline and umbilical veins give rise to the portal venous system and its intrahepatic branches. As fetal development progresses, these early venous connections regress, leading to a complete separation between the systemic and portal circulations. However, the ductus venosus persists, allowing oxygenated blood from the placenta to bypass the liver and flow directly into the inferior vena cava. Within the first few days of life, the cessation of blood flow through the umbilical vein normally triggers the closure of the ductus venosus.[10–12] CPSS arises from incomplete involution of one or more of these embryonic venous structures, resulting in persistent abnormal vascular connections that allow blood to bypass the liver.[12]

Clinical presentations

Clinical presentations of CPSS in pediatric and adult populations are usually highly variable and multisystemic. Some symptoms in CPSS cases can be related to the hemodynamics of the intrahepatic portal veins. This is most conveniently assessed by the size of the intrahepatic portal veins as determined by imaging.[13]

Pediatric presentation

Neonatal cholestasis: Neonatal cholestasis is a common presentation of CPSS, reported by Tran *et al*. in approximately 32% of cases across different cohorts.[14] The underlying pathophysiology is not fully understood, but it is believed to involve either diminished portal flow to the liver or secondary diversion of portal blood due to increased intrahepatic resistance associated with cholestatic liver disease.[15]

In the pediatric population, CPSS are increasingly identified through antenatal or early postnatal ultrasound, either incidentally or as part of newborn screening programs. While many children remain asymptomatic at the time of diagnosis,

others exhibit a broad spectrum of clinical manifestations. The variability in clinical expression is influenced by shunt type, the extent of portal blood diversion, and the timing of diagnosis.[16–19]

Hepatopulmonary syndrome: Hepatopulmonary syndrome has been reported in approximately 3% of patients with CPSS, although higher prevalences of 14–18% have been described in specific cohorts depending on diagnostic definitions and screening methodologies.[8,16,20–22] Hepatopulmonary syndrome results from intrapulmonary vascular dilatations that cause hypoxemia even in the absence of intrinsic liver disease. It can occur at any age, from infancy through adulthood, as a direct consequence of portal blood bypassing the liver and entering the systemic circulation, leading to pulmonary vascular remodeling and impaired oxygenation.[20,23,24] The main clinical presentation includes progressive hypoxemia, cyanosis, and exercise intolerance, particularly during exertion. In severe cases, resting hypoxemia and orthodeoxia (worsening hypoxemia when upright) may be present. Longstanding disease is often associated with digital clubbing and chronic cyanosis. Less frequent manifestations include platypnea (increased dyspnea in the upright position), syncope, and wheezing.[7,25–27]

Portopulmonary Hypertension (PoPH): PoPH is a serious and potentially fatal complication in pediatric CPSS.[5,13,28] The underlying mechanism is attributed to the passage of vasoactive substances and microthrombi into the pulmonary circulation without hepatic clearance, promoting pulmonary vascular remodeling and elevated pulmonary arterial pressures.[23,29] Early clinical manifestations are often subtle, and diagnosis requires a high index of suspicion following the onset of new cardiopulmonary symptoms in children with shunts.[30] Symptoms include dyspnea, cough, syncope, and the appearance of a new right-sided heart murmur.[25,31] Definitive diagnosis is established by right heart catheterization demonstrating elevated mean pulmonary artery pressure with a normal pulmonary capillary wedge pressure, consistent with pre-capillary pulmonary hypertension.[25,32]

Neurocognitive dysfunction and hepatic encephalopathy: Neurocognitive dysfunction in CPSS encompasses a broad spectrum of neurological and psychiatric manifestations, largely attributable to the diversion of portal blood away from the liver, preventing hepatic elimination and leading to systemic accumulation of neurotoxic substances such as ammonia.[5,22,30] These symptoms were reported to range from mild developmental delay and excessive fatigability to learning disabilities[5,33] and, in severe presentations, overt encephalopathy, underscoring the need for early recognition and appropriate treatment.[5,14]

A review of the literature reported that hepatic encephalopathy occurred as a symptom of CPSS in 17–30% of children.[34] In a review of 136 cases of CPSS ranging in age from 0 to 76 years, the prevalence of hepatic encephalopathy (HE) was found to be 13.2%. As the median age of the cases was 6.5 years, it is likely that the majority of the 136 cases were children.[35] Given the risk of progressive neurocognitive impairment, CPSS should be closed as soon as reasonably possible, including during neonatal treatment when appropriate.[4] The likelihood and severity of neurological involvement appear to correlate with the duration of cerebral exposure to ammonia during critical periods of brain development.[35]

Liver nodules/tumors: Liver nodules and tumors are well-recognized presentations of CPSS, occurring in approximately 27.8% of pediatric cases, with a higher prevalence in extrahepatic shunts (EHPSS, 38.2%) compared to intrahepatic shunts (IHPSS, 17.6%).[36] The risk of hepatic neoplasia is particularly elevated in EHPSS, where both benign and malignant lesions have been reported, including hepatocellular carcinoma and hepatoblastoma.[22,36,37] The development of these nodules results from chronic deprivation of portal venous flow, which leads to altered hepatic perfusion and compensatory arterialization, promoting hepatocellular hyperplasia, nodular regeneration, and, in some cases, hepatocellular carcinoma.[36,38–40]

Clinical presentations may vary, ranging from asymptomatic to hepatic dysfunction, most commonly mild elevations in aminotransferases or alkaline phosphatase. Liver nodules are frequently discovered incidentally during investigations for other CPSS-associated complications.[38,39] Benign lesions usually present during childhood or adolescence, while less common malignant lesions can also develop in childhood, including in very young children under the age of five. However, they are more likely to occur after a period of chronic portal deprivation and nodule evolution.[7,17,30]

Adult presentation

In adults, CPSS cases present with a broad and often multisystemic spectrum of manifestations due to chronic diversion of portal blood. The most frequent clinical features include hepatic encephalopathy related to hyperammonemia, pulmonary hypertension, hepatopulmonary syndrome, and the development of benign or malignant hepatic tumors such as focal nodular hyperplasia, adenomas, and hepatocellular carcinoma in the absence of cirrhosis.[5,22,38,41,42] Neurological and psychiatric findings may range from subtle cognitive changes to Parkinsonism, psychiatric manifestations such as psychosis, or recurrent coma following hepatic encephalopathy.[43] In a report of sixty-six mostly adult extrahepatic CPSS patients with a median age of 30 at the end of follow-up, 19 (28%) had HE. The 10-, 20-, and 30-year HE incidence rates were 13%, 24%, and 28%, respectively.[22]

Additional presentations included unexplained hypoxemia, dyspnea, or, rarely, renal involvement such as nephrotic syndrome and glomerulonephritis, likely from systemic exposure to unfiltered metabolites.[44] Despite these complications, liver synthetic function was usually preserved, and portal hypertension was uncommon unless concurrent liver disease was present.[42] Some cases were detected incidentally through abnormal imaging or unexplained laboratory abnormalities such as hyperammonemia, elevated bile acids, or altered galactose metabolism.[42,45] While congenital anomalies, especially cardiac malformations, coexisted in some cases, they were less common in adults compared to children.[5,46] Current guidelines emphasize systematic screening for pulmonary and neurocognitive complications in this population.[41] Baiges *et al*. reported cumulative incidences of 35%, 45%, and 58% of having at least one major CPSS manifestation by the age of 20, 30, and 40 years, respectively.[22]

PoPH has been reported in patients with CPSS, with rates ranging from 7% to 14%. It is the most life-threatening complication of CPSS.[4] Other manifestations, such as liver nodules, may also follow a progressive disease course. The risk of primary HCC in patients with CPSS increases, similar to the risk of HCC in patients with liver cirrhosis. In the former, the shunt appears to work as an independent risk factor for the development of HCC.

Diagnosis of CPSS

CPSS are frequently identified incidentally, often during abdominal or liver imaging conducted for unrelated reasons.[4] In one study, 27% of intrahepatic and 59% of extrahepatic cases were discovered this way. Intrahepatic shunts are typically

asymptomatic at the time of detection and are more commonly diagnosed prenatally, whereas extrahepatic shunts are often identified later in life and tend to be symptomatic.[4,16]

Pre-procedural tests

Accurate visualization and characterization of these shunts are essential for preoperative planning. Because these shunts often lie near vital vessels, precise imaging is necessary to minimize potential complications.[4]

Doppler ultrasonography: A retrospective cohort study by Kivilevitch *et al*. found that intrahepatic portosystemic venous shunts were significantly associated with lower gestational age in fetal growth restriction compared to appropriate gestational age fetuses, as well as an increased risk of preterm delivery, structural abnormalities, and minor genetic aberrations.[47] However, the small sample size of 25 cases limited statistical significance.

Achiron *et al*. reported on 44 cases of fetal umbilical–portal–systemic venous shunts, all diagnosed prenatally using Doppler ultrasound. The study investigated the associated malformations and predictive outcomes of each type. Type I (umbilical–systemic, 20.4%) generally had favorable outcomes, although associated anomalies could have impacted prognosis. Type II (ductus venosus–systemic, 43.2%) presented a more complex range of outcomes influenced by venous system integrity and accompanying malformations. Type III (portal–systemic), further divided into Type IIIa (intrahepatic, 27.2%), had the best prognosis, especially when the intrahepatic portal venous system was intact and no major malformations were present, while Type IIIb (extrahepatic, 9.1%) showed a poorer prognosis, often linked to significant malformations.[48] Doppler ultrasound is recommended as the first diagnostic imaging modality for CPSS in adults. It can detect regenerative nodules and other vascular abnormalities caused by shunting.[15]

MRI and CT: MRI and CT, with and without intravenous contrast, are widely utilized to confirm the diagnosis of CPSS and to provide precise anatomical details of the shunt. Among these modalities, MRI is generally preferred over CT due to its lack of ionizing radiation and superior capability in visualizing hepatic regenerative nodules.[15] Contrast-enhanced MRI with hepatobiliary contrast agents is preferred for preoperative baseline evaluation, as it can detect mild enhancements, increasing its sensitivity for hepatic nodules.

Angiography and occlusion testing: Angiography with temporary occlusion testing is a key component of the preprocedural evaluation in CPSS. This approach involves transiently blocking the shunt to measure portal pressures and assess the capacity of the portal system to accept normal blood flow. Such testing helps predict the ability of the liver to tolerate increased perfusion after closure and guides the prevention of complications like portal hypertension. In general, a portal pressure rise of less than 10 mmHg from baseline during occlusion is considered favorable for single-stage closure. Higher values usually prompt consideration of staged or partial closure to reduce the risk of acute portal hypertension.[49–51]

The test is also indicated for identifying intrahepatic portal branches and larger veins, such as the main portal vein, particularly when these structures are not detected by other diagnostic methods. The test is specifically indicated to detect hypoplastic portal veins and differentiate between end-to-side and side-to-side shunts.[15]

During preoperative assessment, measuring the portosystemic pressure gradient (portal vein pressure – systemic venous pressure, PSPG) is preferred over relying on absolute portal pressure alone because PSPG offers a clearer and more reliable picture of hemodynamic changes. This value more accurately reflects the extra load that shunt closure will place on the portal circulation and helps predict the risk of developing portal hypertension. Because PSPG accounts for variations in both systemic venous and intra-abdominal pressures, it provides a more clinically meaningful measure than absolute portal pressure alone.[52,53]

During the surgical procedure, it is essential to assess the response of the bowel to invasive occlusion testing. Occlusion testing also helps in distinguishing between simple shunts, which typically involve a single communication, and complex shunts, which often feature multiple connections. Recent studies have demonstrated effective vascular access for occlusion testing using the jugular or femoral vein depending on the location of the shunt. In certain cases, dual vascular access may be required, with one access point for occlusion and the other for catheter placement to ensure adequate opacification and accurate pressure measurement.[4,15]

PoPH Screening: Screening for PoPH includes evaluation of patients for shortness of breath, fatigue, and syncope. Transthoracic echocardiography is recommended to estimate pulmonary artery pressure.[4] If high pressures (e.g., >50 mmHg) or right heart dysfunction are found, right heart catheterization is recommended.[54] Screening should be done for both pediatric and adult patients.

Neurocognitive dysfunction tests: Neurocognitive dysfunction tests adapted from HE assessment tools can be helpful for diagnosis and monitoring the effects of treatment on hepatic encephalopathy. A variety of neuropsychological and psychophysiological tools are used to detect minimal HE and related cognitive changes in patients with portosystemic shunting. The psychometric hepatic encephalopathy score, also known as the portosystemic encephalopathy syndrome test, is the most robust and widely validated battery of paper-and-pencil to assess processing speed and visuomotor coordination. Other validated methods include: critical flicker frequency, a non-invasive psychophysiological measure of visual discrimination that correlates with cognitive impairment,[55] and continuous reaction time test, which evaluates motor reaction stability to auditory stimuli and helps distinguish metabolic from organic brain impairment.[56,57] The inhibitory control test, a computerized assessment of response inhibition and working memory, is increasingly applied in pediatric populations.[56] The Stroop test, which measures psychomotor speed and cognitive flexibility and has been validated for minimal HE screening in children with extrahepatic portal vein obstruction.[56,58] Broader neuropsychological batteries encompassing attention, executive function, and fine motor skills (e.g., Grooved Pegboard) are also commonly applied, particularly in pediatric cohorts.[55] These tests are typically complemented by clinical assessment, blood ammonia levels, and, in selected cases, advanced imaging modalities such as magnetic resonance spectroscopy to correlate neurocognitive deficits with metabolic alterations.[5,43,57–60] Screening should be done for both pediatric and adult patients.

Treatment of CPSS

The choice of treatment depends on shunt type, location, degree of function, patient age, and the severity of symptoms and complications.[15] Shunt size and flow are directly related to the likelihood of symptom development.[61] Early intervention is recommended for patients with persistent shunts beyond infancy, symptomatic presentations, or lack of portal vein visualization, as such patients are at increased risk for serious complications.[13,18,19]

A key aspect of preoperative assessment involves deter-

mining (1) whether to pursue an endovascular or surgical approach, and (2) whether closure should be performed in one or two stages.[15,61] In general, long shunts can be closed using endovascular techniques, whereas shorter shunts may be more safely and effectively closed surgically.[4,62] The feasibility of the endovascular approach is determined by two main criteria: first, the occlusion device must not impinge on neighboring vessels, and second, the portosystemic pressure gradient should not exceed 25 mmHg during the occlusion test.[10,15,61] Both endovascular and surgical approaches permit one- or two-stage closures.

Occlusion of end-to-side shunts decreases portal flow to the systemic circulation but does not directly increase intrahepatic portal perfusion. However, the resulting increased portal pressure can indirectly increase intrahepatic portal flow by the formation of collaterals, some of which can empty into the intrahepatic portal system. Occlusion of side-to-side shunts directly decreases portal flow to the systemic circulation and increases intrahepatic portal perfusion. A multidisciplinary approach should be utilized to manage systemic manifestations and the development of further complications.[62] Treatment of late CPSS is often complicated and requires careful evaluation of hepatic and renal function and hepatic blood flow. Because of pre-existing CPSS, gastrointestinal bleeding is best managed by medical and endoscopic measures to preserve hepatic perfusion.[2]

Endovascular treatments

Endovascular techniques for the treatment of CPSS are minimally invasive procedures performed by interventional radiologists to occlude abnormal vascular communication between the portal and systemic venous systems.[48,63] Endovascular techniques are considered the first choice for treatment of CPSS, as they are associated with shorter procedure times, less blood loss, and more favorable outcomes compared to surgical ligation.[64]

Transcatheter embolization: This is the most common approach and involves insertion of coils, vascular plugs, or microvascular plugs to achieve shunt occlusion. The choice of device depends on shunt size, length, and flow characteristics. Coils are typically used for small, narrow shunts, while vascular plugs are preferred for larger or high-flow shunts.[9,65]

Staged endovascular closure: In patients with hypoplastic portal veins or elevated portal pressures, staged reduction of shunt flow may be performed using a reducing stent or partial occlusion with a modified plug, followed by delayed complete closure after portal vein growth and pressure normalization.[51]

Knirsch *et al*. reported a case series of eight children with congenital portosystemic venous shunts managed through catheter-based interventions.[24] Diagnostic evaluation included balloon occlusion testing and angiography to assess portal vein development. Interventions ranged from partial to complete shunt closure using vascular plugs and coils. All eight procedures were technically successful with no major complications reported. Five patients underwent shunt closure at a median age of 3.9 years (range: 0.7–21 years), while three patients were not treated due to clinical stability, palliative status, or future procedural planning. Among the treated group, follow-up demonstrated significant portal vein growth confirmed by catheterization in cases of partial closure and by ultrasound in cases of complete closure. Portal vein caliber and flow improved in patients with initially rudimentary or small intrahepatic portal veins, supporting the effectiveness of endovascular treatment in promoting vascular remodeling.[24] However, the small sample size, single-center experience, and retrospective design limit the generalizability of the findings.

Zhang *et al*. conducted a retrospective study comparing surgical ligation and endovascular embolization for Type II congenital extrahepatic portosystemic shunts, demonstrating that both approaches were effective and safe, with clinical improvement and normalization of ammonia levels observed in all 23 patients within 6–12 months post-procedure.[66] Endovascular embolization was associated with significantly shorter procedure times, less intraoperative blood loss, and favorable portal vein remodeling, with a significant increase in portal vein diameters. However, surgical ligation remained a valuable alternative for patients with short, broad shunts or elevated portal pressures, particularly when combined with splenic vessel ligation. In the surgical group, post-procedural portal vein pressure increased significantly, although remaining below 25 mmHg. Clinical symptoms such as hepatic encephalopathy and gastrointestinal bleeding resolved in both groups, with only one case of rebleeding (gastric ulcer) and one case of portal vein thrombosis, which was managed successfully. The choice of intervention should be individualized based on anatomical considerations and portal hemodynamics.[66] The study was limited by its small sample size, retrospective design, and absence of standardized follow-up intervals and portal pressure gradient measurements, which may restrict the generalizability of the findings.

In general, endovascular approaches are preferred for long, narrow shunts, while surgical intervention may be required for short and broad shunts or when catheter-based access is not technically feasible. However, data on long-term outcomes and optimal treatment strategies for complex or atypical shunt anatomies remain limited, highlighting the need for continued longitudinal follow-up and collaborative experience sharing.

Surgical treatment

The Bicêtre surgical classification categorizes CPSS into four distinct types based on their anatomical configuration and the termination of the shunt within the caval system.[67] This classification provides a structured framework that aids in determining the most appropriate surgical or interventional approach.[5,67] Extrahepatic shunts can be closed in one step (either by interventional radiology or surgery), Table 1. The Abernethy type I (end-to-side retrohepatic portocaval shunt) is frequently closed in two steps,[10,67–70] but may be closed in one step to avoid the development of portal hypertension, while the Abernethy type II (side-to-side retrohepatic portocaval shunt) can be closed in one step, Figure 1.[28,68]

Some cases warrant special observation and monitoring.[71] Intrahepatic shunts diagnosed during infancy or prenatally may close spontaneously by one year of age with resolution of symptoms.[9,15,61,72] The treatment of asymptomatic CPSS before the first year of life is controversial, and data are largely limited to case reports. In contrast, it is recommended that all extrahepatic or persistent intrahepatic shunts beyond the first year of life be closed.[9,15,61,72] Additionally, the presence of clinical encephalopathy, hepatopulmonary syndrome, PoPH, liver lesions, and evidence of increasing shunt size are all clear indications for intervention.[9,15,61,72] It has been proposed that even in the absence of overt symptoms, early intervention can prevent life-threatening cardiopulmonary and neurological complications.[15,49,61] Given the retrospective nature of existing studies, small sample sizes, and short follow-up periods, these studies are limited in their generalizability.

Closure of portosystemic shunts allows subsequent growth of the portal vascular system, thus preventing or reversing associated signs and symptoms.[73] Preoperative assess-

Table 1. Surgical Treatment of CPSS by Subtype

CPSS subtype	Key features	Typical surgical approach	Radiological features	Notes
EHPSS, PH fistulas (superficial), PDV	Adequate intrahepatic portal system	Single-stage ligation	N/A	Usually, a well-developed portal system allows direct closure[67]
ESPCS	Thread-like or absent IPVS, poor bowel tolerance on occlusion	Two-stage: initial banding → delayed closure	Uniform PV draining into left IVC, absent intrahepatic portal branches (IHPB)[68]	Risk of portal hypertension if closed at once; spontaneous closure may occur after banding[67–69]
SSPCS	Patent IPVS, good bowel tolerance on occlusion	One-stage caval partition or ligation	Aneurysmal PV draining anteriorly/rightward into IVC with visible IHPB[70]	Favorable anatomy for single-stage closure; spontaneous closure also reported[10,67–69]
All subtypes (if hepatoportal flow restored)	Restoration of portal flow may resolve symptoms	Secondary closure sometimes unnecessary	Depends on subtype	Spontaneous closure post-banding observed in both ESPCS and SSPCS, supporting conservative treatment[68,69]

CPSS, congenital portosystemic shunts; IHPSS, intrahepatic portosystemic shunts; EHPSS, extrahepatic portosystemic shunts; IVC, inferior vena cava; HCC hepatocellular carcinoma; PPH, portopulmonary hypertension; PV, portal vein, N/A, not applicable; PH, portal hypertension; PDV, patent ductus venosus; ESPCS, end-to-side portocaval shunts; SSPCS, side-to-side portocaval shunts; IPVS, intrahepatic portal venous shunts; IHPB, intrahepatic portal branches.

ment should aim to define the shunt anatomy, pressures, and flow. This ensures a safe and personalized approach and helps mitigate procedural risks. Careful treatment of extrahepatic manifestations should be done before shunt closure. Medical therapy is utilized at this stage. For CPSS associated with PoPH, endothelin receptor antagonists, phosphodiesterase-type 5 inhibitors, and prostacyclin analogues have been used to manage systemic disease and improve surgical outcomes.[10] A systematic review conducted by Galie *et al*., which included patients with pulmonary arterial hypertension demonstrated that medical therapy resulted in improved exercise capacity, hemodynamics, and outcomes compared with untreated patients.[69] However, variability in trial design, such as differences in patient populations, background therapies, trial endpoints, and short follow-up periods, limits the generalizability of the findings.

Uike *et al*. described 24 patients with CPSS, of whom 54% had extrahepatic, 20% portocaval, 17% portohepatic, and 8% persistent ductus venosus and nine patients diagnosed-with pulmonary hypertension. Five of these patients underwent closure, and postoperative follow-up showed improvement of PoPH without complete resolution. PAH-specific drugs given in conjunction with CPSS closure resulted in greater improvement in pulmonary portal hypertension hypertension and right ventricular pressure compared to medical therapy alone.[28] The study was limited by the small sample size and short follow-up period.

Uchida reported on 55 patients diagnosed with congenital extrahepatic portosystemic shunts, 44 of whom were managed by endovascular closure, surgical closure, or liver transplantation. Reported postoperative complications included splenomesenteric vein thrombosis, portal hypertension, and progression of PoPH.[68] The findings underscored the importance of postoperative monitoring and additional treatment in some patients who undergo shunt closure. The small sample size and retrospective nature of the study are limitations.

Zhang *et al*. reported on 12 patients with CPSS treated with surgical ligation because of (1) a positive occlusion test and (2) a lack of experience in endovascular closure by the treating institution. Six patients underwent single-stage ligation of the shunt, five underwent two-stage ligation, and one was treated with partial ligation. All patients experienced resolution of hyperammonemia postoperatively and had satisfactory outcomes. Postoperative thrombosis specifically at the ligation site was a specific concern.[66] This study highlighted the importance of postoperative monitoring and preventive treatment with anticoagulation. The small number of patients, short follow-up period, and unclear selection criteria for the surgical approach were limitations.

Mori *et al*. described two cases of laparoscopic partial closure for extrahepatic CPSS in which the occlusion test was positive for elevated portal vein pressure exceeding 25 mmHg. Partial closure alone was performed in the first case, while a staged approach was undertaken for the second patient, with complete closure performed six months following the initial intervention.[49] In all cases, the patients demonstrated improvement in laboratory markers and showed no signs of liver dysfunction, encephalopathy, or portal hypertension following closure. The study was hampered by a small sample size, short follow-up period, and limited reporting on postoperative treatment, including the criteria for complete closure. Although there is currently no official treatment guideline, a portal pressure threshold of 25–32 mmHg has been recommended in the literature. Tran *et al*. recently recommended a cutoff of 30 mmHg.[14]

For liver tumors associated with intrahepatic or extrahepatic CPSS, it is recommended to close any shunt regardless of patient age.[49,74] It should be noted that the behavior of the tumor following closure can be unpredictable. Closing the shunt may lead to partial or complete regression of the mass by restoring normal arterial and portal flows.[49] In cases of partial regression, reassessment of vasculature is essential before surgical resection.[49] In some cases, nodule resection may be performed concurrently with shunt closure.[49] Malignant masses require standard oncological treatment in addition to shunt closure.[49] Caution should be used with embolization of HCC, as it is associated with significant ischemic liver injury.[61] Franchi-Abella *et al*. reported a series of 22 CPSS patients presenting with single or multiple benign and malignant liver lesions. Following shunt closure, partial regression was observed in three patients, while complete regression was seen in seven patients.[2] For malignant tumors, tumor resection was performed concurrently with shunt closure. Similarly, Grimaldi *et al*. described a case of a child with hepatopulmonary syndrome and a liver mass that was managed by radiologic intervention. Following closure of the

Table 2. Recommended Surgical Approaches for CPSS Indications

Surgical approach	Recommended indications
Liver resection	Large, multifocal intrahepatic shunts obstructing or malignant liver tumors, shunts rapidly increasing in size or changing features[61]
Liver transplantation	Type 1 extrahepatic CPSS with failed occlusion test,[61,75] severe underlying liver disease,[20,22,69] multifocal or growing nodules with biopsy-proven malignancy,[20,61,69] severe portal hypertension[10,11,61,69]
Either approach considered	Failed radiological intervention,[2,10,61] development of collateral vessels after shunt closure[69]

CPSS, congenital portosystemic shunts.

shunt, the patient showed improvement and partial regression of the mass at the 3.5-month follow-up.[75] Liver resection or transplantation is generally recommended as a last resort in the treatment of CPSS. Table 2 describes indications for liver resection and transplantation.[2,10,11,20,22,61,69,75]

Uchida *et al*. evaluated clinical data and outcomes of extrahepatic CPSS in 29 patients who underwent liver transplantation. Nineteen percent of patients developed surgical complications, including biliary complications, vascular complications, intra-abdominal hemorrhage, infections, and immunosuppressant-related complications.[76] All patients demonstrated clinical improvement or lack of progression of preoperative CPSS-related complications. The retrospective nature of the study, small sample size, and variations in follow-up periods are notable limitations.

Conclusions

CPSS are rare vascular anomalies that allow portal blood to bypass liver detoxification, leading to serious complications. While animal studies suggest a genetic basis, the cause in humans remains unclear, although associations with other anomalies point to a genetic role. CPSS clinical manifestations include hepatic encephalopathy related to hyperammonemia, PoPH, hepatopulmonary syndrome, and the development of benign or malignant hepatic tumors such as focal nodular hyperplasia, adenomas, and hepatocellular carcinoma. Recent publications recommend closure of asymptomatic intrahepatic CPSS if they do not close spontaneously within two years of age, and all asymptomatic extrahepatic CPSS as early as possible. This is to prevent sequelae of chronic hepatic hypoperfusion, fibrosis, and tumor formation. Treatment strategies for CPSS depend on shunt size, occlusion test results, tumor presence, and the early or late stage of disease. Our findings support screening for CPSS in patients, especially young individuals, who present with unexplained hepatic encephalopathy in the absence of cirrhosis or portal hypertension.

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Conflict of interest

GYW has been an editor-in-chief of *Journal of Clinical and Translational Hepatology* since 2013. He had no role in the review process of this manuscript.

Author contributions

Proposed the concept for the review, drafted the article, revised the manuscript critically, and approved the final version and publication of the manuscript (GYW).

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