

Status of congenital extrahepatic portosystemic shunt

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To the Editor: Studies have demonstrated that congenital extrahepatic portosystemic shunt (CEPS) is a serious and life-threatening disease.^[1] According to statistics, deformity, infection, liver failure, and failure of other systems are the main causes of death.^[2] This article will introduce the pathology, classification, clinical manifestations, diagnosis, and treatment of CEPS.

Type I CEPS is caused by excessive degeneration of the vitelline vein ring around the duodenum or failure of the vitelline vein to anastomose with the hepatic sinus or umbilical vein, resulting in the persistence of the right vitelline vein (shunt to the posthepatic inferior vena cava (IVC) or the left vitelline vein (shunt to the IVC or the right atrium is higher than the confluence level of the hepatic vein)).^[3] The development of embryonic IVC is also very complicated. The vitelline vein is anastomosed with the IVC, which developed from the IVC near PV (portal vein); if this mild channel persists, it will cause type II CEPS.

Currently, the most prevalent classification of CEPS is that Morgan *et al* classified it into type I (absent PV), that is, an end to side shunt type and type II (hypoplastic PV), that is, a side-to-side shunt type based on the presence or absence of PVs. Type I was further divided into type Ia: superior mesenteric vein (SMV) and splenic vein (SV) were drained into systemic vein, respectively; Type Ib: SMV and SV confluence to form a common trunk and then discharge into systemic vein [Figure 1]. With the continuous discovery of CEPS, people put forward different classification methods. According to the location of portal blood flow, CEPS is divided into: (A) SMV and SV into IVC, (B) SMV and SV into the left renal vein, and (C) SMV and SV into the iliac vein; according to the source of shunt entrance, type II CEPS is classified into type IIa (derived from PV branches), type IIb (derived from PV trunk, bifurcation, or spleen–intestinal confluence), and type IIc (derived from the mesenteric vein, gastric vein, or SV); according to the end of shunt cavity, CEPS is divided into extrahepatic portosystemic shunt, portal cava, portohe-

patic, persistent ductus venosus; according to the degree of intrahepatic portal venous system dysplasia, CEPS is classified into mild, moderate, and severe.

The clinical presentation of CEPS is diverse and can even appear asymptomatic until old age or late in the course of the disease.^[4] The clinical manifestations of CEPS are mainly divided into three categories: lesions caused by insufficient hepatic blood perfusion, symptoms caused by the extrahepatic portal shunt, and malformations associated with other systems. The reduced blood supply to the liver can lead to hepatic atrophy, hepatic steatosis, and liver nodules. Among them, liver nodules occur in up to 65%,^[2] and although most of them are benign, they have a high risk of malignant transformation. This may be related to hepatic arterial hypertension, PV ischemia, and hepatocyte growth factor. Systemic shunting of splanchnic venous return can lead to abnormal liver development, regeneration, and function.

Hepatopulmonary syndrome, pulmonary hypertension, and hepatic encephalopathy are the most prominent manifestations caused by long-term portosystemic shunting. Pulmonary hypertension has a poor prognosis and cannot be reversed even after late closure of the shunt, with a mortality rate of 50%.^[1] The incidence of encephalopathy increases with age. Other neurological complications, such as hepatomyelopathy, parkinsonism, and pyramidal syndrome, have also been reported. CEPS can cause mesenteric circulation, avoiding associated harmful compounds (ammonia, intestinal vasoactive mediators, lactate, etc.) produced during digestion.^[5] In addition, it may cause gastrointestinal bleeding, hormonal disorders, endocrine disorders, anemia, and peripheral edema. Cardiovascular malformations are the most common congenital defects in CEPS. Genitourinary malformations, digestive system malformations, musculoskeletal defects, and other conditions (cutaneous hemangiomas, Goldhar syndrome, Caroli syndrome, Turner syndrome, and Down syndrome) have also been reported.

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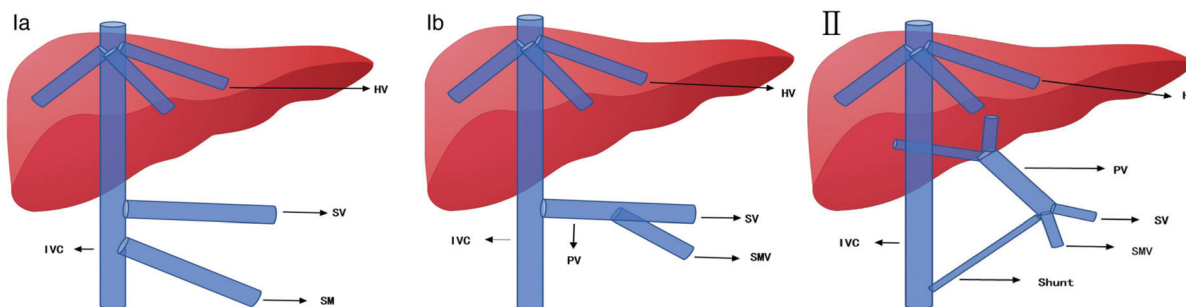


Figure 1: The classification of CEPS. Type I (absent PV) and type II (hypoplastic PV). Type I was further divided into type Ia: SMV and SV were drained into systemic vein, respectively; Type Ib: SMV and SV confluence to form a common trunk and then discharge into systemic vein. CEPS: Congenital extrahepatic portosystemic shunt; HV: Hepatic vein; IVC: Inferior vena cava; PV: Portal vein; SMV: Superior mesenteric vein; SV: Splenic vein.

Because of the lack of specificity in the biological and clinical manifestations of CEPS, ancillary examinations, especially imaging examinations, are essential for the diagnosis, classification, and screening of related complications of CEPS. Clinically, appropriate examination methods should be selected according to the age of patients, shunt anatomy, clinical symptoms and complications, and the possibility of concurrent treatment. Doppler ultrasound can show the abnormal relationship between PV and other veins, liver morphology, liver nodules, and other malformations, and it is noninvasive, economic, and practical, which can be used as a preliminary screening of CEPS, especially in prenatal screening. In addition, enhanced color Doppler ultrasound can also be used to estimate the shunt ratio (the blood flow at the shunt divided by the total flow of the PV). Magnetic resonance imaging (MRI) is the preferred examination for CEPS. MRI is ionizing radiation free, and assessment of hepatic vascular anatomy is reliable and noninvasive, with good performance in detecting associated focal hepatic lesions, parenchymal hepatic lesions. In addition, brain injury due to shunting can also be shown to manifest as white matter atrophy or globus pallidus hyperintensity on T1 weighted images.^[6] In detecting small vessel branches, computed tomography angiography (CTA) has a higher spatial resolution than MR imaging, in addition, CT can image all abnormal vessels at one breath hold and can be used in patients who are not operated on or have specific contraindications.^[7] Other imaging examinations help find complications of CEPS. Bubble echocardiography or scintigraphy to detect intrapulmonary arteriovenous shunts; echocardiography to screen for pulmonary hypertension; and cardiopulmonary catheterization to screen for indirect signs of pulmonary hypertension. Pulse oximetry, contrast-enhanced ultrasound, and pulmonary perfusion imaging are helpful in the diagnosis of hepatopulmonary syndrome. Clinically, many patients are diagnosed because of complications of CEPS or other reasons requiring biopsy or resection of liver specimens. Liver biopsy can find the changes of PV and its branches in different degrees. However, PV dysplasia is easily diagnosed as type I CEPS on pathological examination and is more subjectively affected by the operator. Angiography is the reference standard for the diagnosis of CEPS and it is particularly suitable for extrahepatic portosystemic shunts and end-to-side portosystemic shunts, for finding severely dysplastic and possibly heterotopic PVs and their branches, and for assessing PV

pressure during shunt occlusion. Besides, portography can also help measure pressure gradients, indicate venous flow, perform interventional embolization, and help identify vascular tumors and display the results of MRI. Owing to its shortcomings of radiation exposure, the need for anesthesia and vascular puncture, and concomitant complications, it is generally not the preferred test for the diagnosis of CEPS. However, the results proved to be relatively safe. Angiography and occlusion tests are essential when intrahepatic PV branches and major PV are not visible. Many patients with CEPS are classified as type I by CT or MRI, but further studies of intravascular shunt occlusion tests have revealed that there are residual hypoplastic veins.^[1,5,7] The most important for the differentiation of CEPS is to distinguish between types I and II. CTA and MR images are helpful in differential diagnosis. If type I CEPS is diagnosed by imaging, angiographic occlusion test is necessary.

There is currently no consensus on the treatment options for CEPS. Specific treatment options should be selected according to the type of deformity and the specific circumstances of the patient. Current treatment options for CEPS include surgical or interventional correction of shunts, hepatic nodule resection, and liver transplantation. Liver transplantation is the only effective treatment for patients with type I CEPS. However, shunt blockade is ineffective, because shunts represent the only drainage route for mesenteric and splenic venous blood. The most common methods of the closed shunt in patients with type II CEPS are interventional closure and surgical ligation, of which interventional occlusion is the preferred treatment for type II CEPS, and it includes balloon occluded retrograde transvenous occlusion and metallic coil embolization. When placing an occlusive balloon, consideration should be given to the location of the systemic venous connection that is long enough to accommodate the occlusive balloon (patented venous catheter or other communication).^[7] Measuring PV pressure during the occlusion test can determine the closure pattern. In well-tolerated patients, shunt block may be attempted, but elevated portal pressure may require two-stage surgery. That is, choroids are first formed through the shunt, and then the shunt is closed. It has been reported that if PV pressure is 25 to 30 mmHg or higher after balloon occlusion, it is difficult to close shunt. If the portal pressure is 32 mmHg or higher, a two-stage shunt (closing the shunt

3–10 months after the first treatment) can be used. Percutaneous occlusion is usually performed as long as the occluder can be fixed in the shunt without affecting the venous drainage position. Surgical closure of the shunt is preferred if the patient is not suitable for intraluminal closure or if the shunt is wide and short.^[4] When the PV is severely hypoplastic, shunt ligation needs to be performed in two steps. Portal pressure was measured during initial clamp shunting and intestinal congestion was observed. If the portal pressure exceeds 32 mmHg after shunt occlusion, the pressure needs to be controlled around 20 mmHg for several months before final closure occurs. There is literature suggesting that surgical occlusion may increase the risk of neovascular shunting. In addition, for patients with a low portosystemic shunt and PV plasticity, PV thrombosis may result postoperatively. Shunt closure must always be considered in patients with symptomatic CEPS. Shunt closure can be used as a preventive treatment early in the disease progression to prevent the occurrence of serious complications. For hepatic encephalopathy, shunt closure is particularly effective.

However, the risk of complications cannot be reduced in some patients after shunt closure,^[1] such as combined severe pulmonary hypertension. It may also cause complications after shunt closure, such as increased liver enzyme levels, thrombosis, the appearance of a new shunt, the recurrence of portal hypertension, and the recurrence of hyperammonemia. Therefore, long-term follow-up is needed to monitor shunt complications, particularly portal hypertension and the appearance of new shunts after shunt closure. Controversy exists regarding hepatic nodular lesions in patients with CEPS. Its hepatic nodular lesions are mostly benign, and some nodules shrink or regress after shunt closure, suggesting long-term monitoring and follow-up. However, its malignant transformation rate was high, and there were cases in which malignant lesions developed during follow-up.

CEPS is a rare congenital disorder, and most patients die of severe shunt-related morbidity. Because of the complex presentation of CEPS, when there is an unexplained

associated disease, one should be alert to the possibility of CEPS and recommend CT or MRI imaging to confirm the diagnosis. Doppler ultrasound is useful in aiding diagnosis in infants. If a diagnosis of type I CEPS is made, an angiographic occlusion test is recommended for further definitive classification, and liver transplantation is preferred in patients with type I CEPS; type II CEP should be selected for shunt closure. Patients with CEPS should be carefully monitored for complications and shunting in the long term postoperatively.

Conflicts of interest

None.

References

1. Baiges A, Turon F, Simón-Talero M, Tasayco S, Bueno J, Zekrini K, *et al.* Congenital extrahepatic portosystemic shunts (Abernethy Malformation): an international observational study. *Hepatology* 2020; 71:1–12. doi: 10.1002/hep.30817.
2. Kobayashi N, Niwa T, Kirikoshi H, Fujita K, Yoneda M, Saito S, *et al.* Clinical classification of congenital extrahepatic portosystemic shunts. *Hepatol Res* 2010; 40:585–593. doi: 10.1111/j.1872-034X.2010.00667.x.
3. Sanada Y, Mizuta K. Congenital absence of the portal vein: translated version. *J Hepatobiliary Pancreat Sci* 2018; 25:359–369. doi: 10.1002/jhbp.572.
4. Papamichail M, Pizaniyas M, Heaton N. Congenital portosystemic venous shunt. *Eur J Pediatr* 2017; 177:285–294. doi: 10.1007/s00431-017-3058-x.
5. Jain V, Sangdup T, Agarwala S, Bishoi AK, Chauhan S, Dhua A, *et al.* Abernethy malformation type 2: varied presentation, management and outcome. *J Pediatr Surg* 2018; 54:760–765. doi: 10.1016/j.jpedsurg.2018.08.053.
6. Pujol A, Pujol J, Graus F, Rimola A, Peri J, Mercader JM, *et al.* Hyperintense globus pallidus on T1-weighted MRI in cirrhotic patients is associated with severity of liver failure. *Neurology* 1993; 43:65–69. doi: 10.1212/wnl.43.1part1.65.
7. Franchi-Abella S, Gonzales E, Ackermann O, Branchereau S, Pariente D, Guérin F, *et al.* Congenital portosystemic shunts: diagnosis and treatment. *Abdom Radiol (NY)* 2018; 43:2023–2036. doi: 10.1007/s00261-018-1619-8.

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