Revascularization of Portal Venous System after Occlusion of Congenital Intrahepatic Portosystemic Shunt

An 11-month-old boy was admitted to our hospital due to unremitting hyperammonemia. He was diagnosed as having an intrahepatic portosystemic shunt (IPSS) at 24 weeks of gestation. He had no significant symptoms, and physical examination revealed good growth and development. Laboratory results showed an elevated serum ammonia level of 61.6 μmol/L (normal, 0-33 μmol/L), along with normal serum bilirubin and aminotransferase levels.

Abdominal ultrasonography revealed a fistula between the portal vein and the inferior vena cava. Computed tomography angiography (CTA) of the abdomen demonstrated the portal vein blood directly draining into the inferior vena cava via a shunt located in the right lobe of the liver; the intrahepatic portal branches could not be visualized (**Figure 1**). After evaluating the risks and benefits, we opted for interventional therapy.

Portography further confirmed the large-sized shunt and hypoplastic portal branches (**Figure 2**). When the balloon temporarily blocked the shunt, the portal venous pressure was measured at 16 mmHg. Then a 12/14-mm vascular plug was selected to inserted into the proper position to ensure the development of the portal system and the confluence of the hepatic vein. The patient's blood ammonia levels fell to normal in 3 days, and the hypoplastic intrahepatic portal branches were visualized by CTA after 2 months (**Figure 3**).

Congenital IPSS is an uncommon anomaly in which the portal vein blood flows directly into the systemic circulation.[1] In early infancy, children may present with hypergalactosemia or hyperammonemia; however, if the shunt persists, patients can manifest hepatic encephalopathy, pulmonary hypertension, or liver tumors.[2] Occluding the anomalous shunt and restoring the liver flow is the basic therapeutic principle; however, available data suggest that the treatment should be delayed because spontaneous closure can be expected in the first 2 years of life if no complications are detected or if the shunt ratio is <30%.[3] Although our patient was only 11 months old and asymptomatic, we chose to intervene because chronic hyperammonemia may have a harmful influence on the child's developing brain.[2] Previous research suggested that the hypoplastic portal system can develop through a cavernomatous hepatopetal network after closure of the shunt.[4] Kanazawa et al also indicated that treatment should be performed as soon as possible; if delayed,

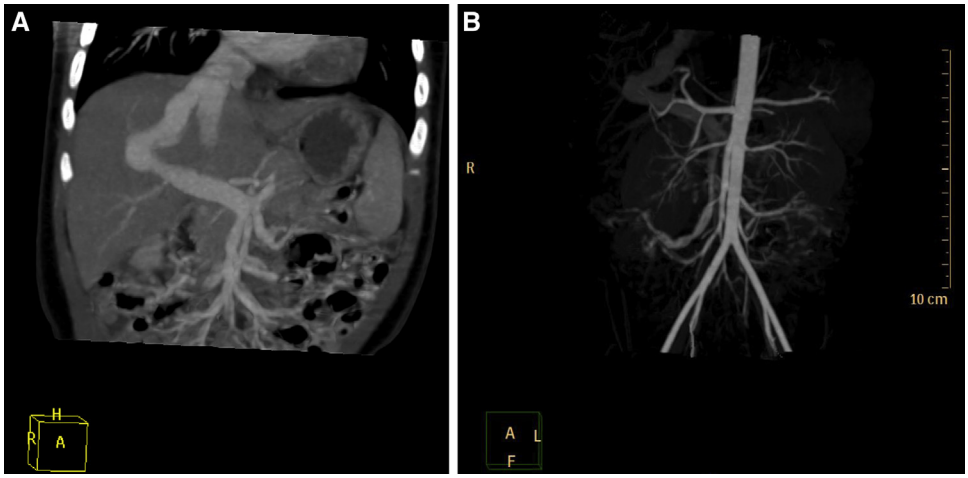

Figure 1. CTA of the abdomen showing hypoplastic intrahepatic portal branches.

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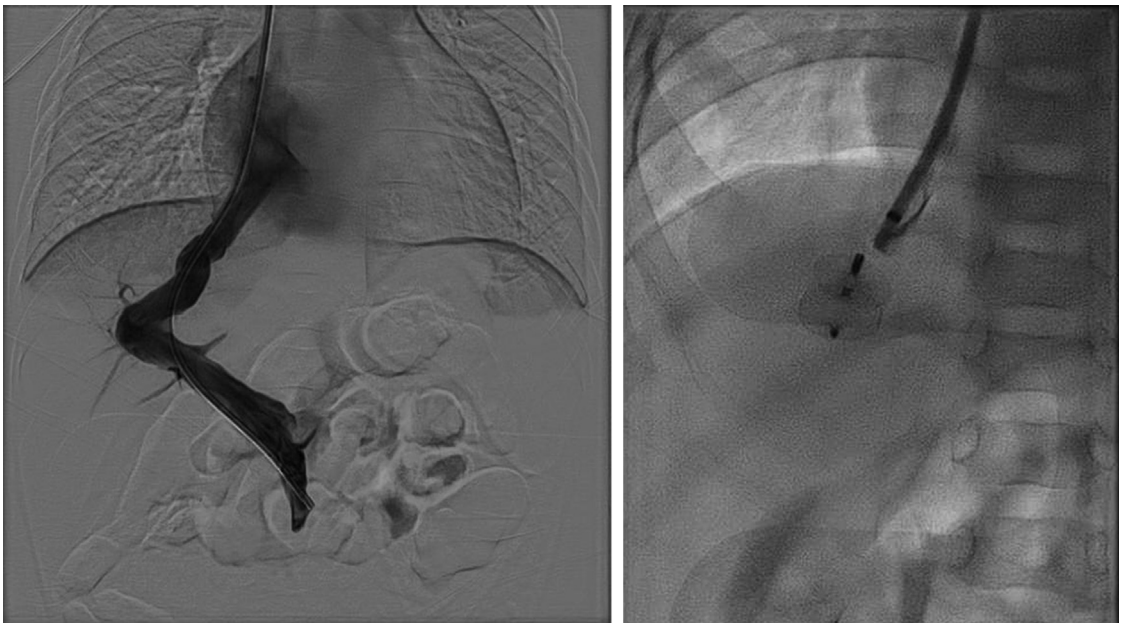

Figure 2. **A,** Portography before vascular plug placement indicating an IPSS and hypoplastic portal branches. **B,** The position of the plug confirmed by portography after the operation.

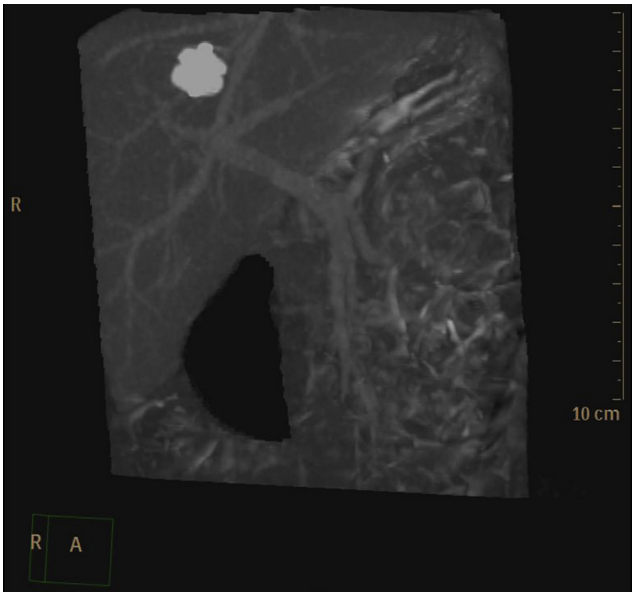

Figure 3. Follow-up CTA at 6 months postoperation demonstrating revascularization of portal venous system.

closure may be more difficult as the shunt grows with age.[5] Furthermore, few cases of severe complications related to shunt closure have been reported to date. ■

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References

1. Stringer MD. The clinical anatomy of congenital portosystemic venous shunts. Clin Anat 2008;21:147-57.
2. Bernard O, Franchi-Abella S, Branchereau S, Pariente D, Gauthier F, Jacquemin E. Congenital portosystemic shunts in children: recognition, evaluation, and management. Semin Liver Dis 2012;32:273-87.
3. Palvanov A, Marder RL, Siegel D. Asymptomatic intrahepatic portosystemic venous shunt: to treat or not to treat? Int J Angiol 2016;25:193-8.
4. Franchi-Abella S, Branchereau S, Lambert V, Fabre M, Steimberg C, Losay J, et al. Complications of congenital portosystemic shunts in children: therapeutic options and outcomes. J Pediatr Gastroenterol Nutr 2010;51:322-30.
5. Kanazawa H, Nosaka S, Miyazaki O, Sakamoto S, Fukuda A, Shigeta T, et al. The classification based on intrahepatic portal system for congenital portosystemic shunts. J Pediatr Surg 2015;50:688-95.