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Successful Surgical Ligation Under Intraoperative Portal Vein Pressure Monitoring of a Large Portosystemic Shunt Presenting as an Intrapulmonary Shunt: Report of a Case

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Abstract
We report a rare case of patent ductus venosus (PDV) with collapsed intrahepatic portal branches and an intrapulmonary shunt. Excellent improvement of the intrahepatic portal vein flow was achieved by ligating the large ductus venosus under intraoperative portal vein pressure (PVP) monitoring. A 3-year-old boy being followed up for hypergalactosemia at a local hospital was found to have mild lip cyanosis, exertional dyspnea, clubbed fingers, and mild liver dysfunction with high levels of transaminase and ammonia. Cardiac catheterization indicated an intrapulmonary shunt with a ratio of 33%. Abdominal ultrasonography and computed tomography showed remarkable communication between the portal vein and the inferior vena cava. We performed laparotomy and successfully ligated the PDV under PVP monitoring. The PVP did not increase until the catheter was removed 7 days postoperatively. The patient's liver function test results returned to normal within 2 weeks. His serum galactose level was 0 mg/dl after drinking milk, and his SpO_2 in room air and exertional dyspnea also improved. He was discharged 18 days after his operation, without any complications. We propose that ligation of a PDV under PVP monitoring could be a treatment of choice, bearing in mind that PDV is associated with collapsed intrahepatic portal branches.

Key words Patent ductus venosus · Portal vein pressure monitoring · Ligation · Intrapulmonary shunt

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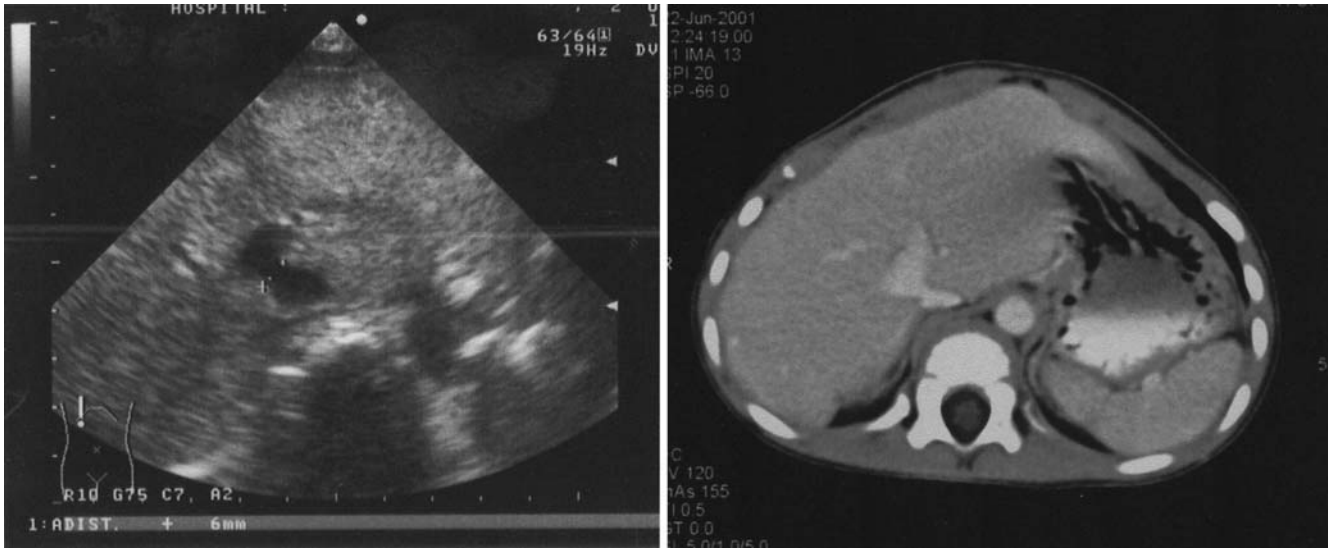
Introduction

The patent ductus venosus (PDV) is a rare form of congenital portosystemic shunt, characterized by the same signs and symptoms as a portocaval shunt. Its clinical features have not been clarified, and there have been few reports of surgical treatment for PDV in children. The reported treatments include liver transplantation, banding, coiling, or stenting,[1] but there is no standard operation for symptomatic PDV. To our knowledge, this is the first pediatric case of PDV with collapsed intrahepatic portal branches and an intrapulmonary shunt, in which excellent improvement was achieved by ligation under intraoperative portal vein pressure (PVP) monitoring.

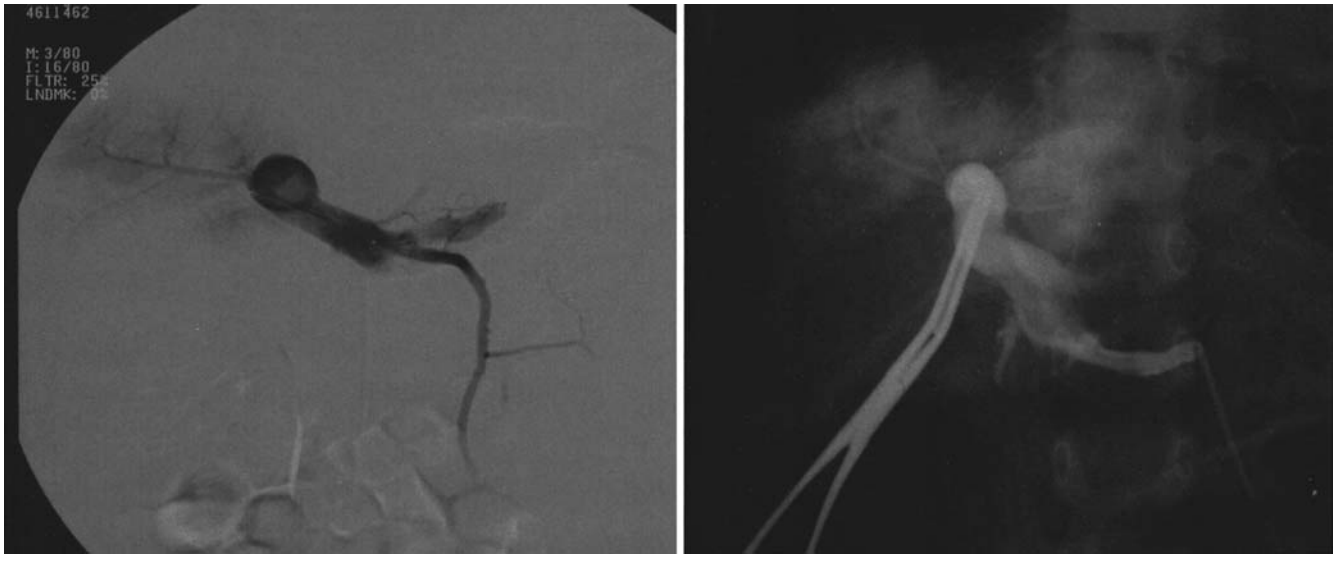
Case Report

A 3-year-old boy being followed up for hypergalactosemia at a local hospital was found to have mild lip cyanosis, exertional dyspnea, clubbed fingers, and mild liver dysfunction with high transaminase and ammonia levels. A cardiac catheterization was done for the unexplained cyanosis, which suggested an intrapulmonary shunt without pulmonary hypertension. An open liver biopsy showed no hypoplasia of the intrahepatic portal vein or fibrosis, implying that liver transplantation was not indicated. The patient was subsequently referred to our hospital for surgical management. The child was mildly intellectually delayed, with a development quotient of 58 points. Laboratory examinations showed slightly increased levels of serum ammonia (85 μg/dl), total bilirubin (1.8 mg/dl), aspartate aminotransferase (58 IU/l), and serum bile acid (62 μmol/l), with additional mild coagulopathy (prothrombin time 17.1 s). The portosystemic shunt ratio was estimated at 75% by portal scintigraphy using [^{123}I]-iodoamphetamine, administered rectally. In room air, SpO_2 was 86% and blood gas



A B

Fig. 1. Preoperative ultrasonography (**A**) and computed tomography (**B**) showed a shunt between the portal vein and the inferior vena cava



A B

Fig. 2. **A** Preoperative portography showed collapsed intrahepatic portal vein branches with a balloon blockade of the ductus venosus. No left branches were seen. **B** Intraoperative portography showed left branches after the ductus venosus was clamped

analysis showed 61 torr. The intrapulmonary shunt ratio was 33% on ^{99m}TcMAA scintigraphy.

Abdominal ultrasonography and computed tomography showed a large communication between the portal vein and the inferior vena cava (Fig. 1), but there was no hypoplasia of the hepatic vein. Preoperative portography also showed collapsed intrahepatic portal branches (Fig. 2A).

We initially considered that surgical intervention was impossible, because of the size of the shunt. Histology did not show any hypoplasia of the intrahepatic portal vein or fibrosis, based on which live transplantation was not thought to be a valid option. Thus, we decided to perform either ligation or banding under intraoperative PVP monitoring, to prevent any sudden increase in the PVP.

Laparotomy revealed a soft liver with a smooth surface and no apparent atrophy or ascites. To measure the PVP, we introduced a BioLine Catheter (Nipro, Osaka, Japan) from the inferior mesenteric vein to the portal

vein. We found a large shunt, about 8 mm in diameter (Fig. 3). After temporary test clamping of the PDV, the PVP increased from 5 to 13 mmHg, but it was still within normal limits and showed no change for 20 min (Fig. 4). There was no congestion of the small intestine. Intraoperative portography showed the left branches of the intrahepatic portal vein (Fig. 2B). Based on these findings, we decided to ligate the PDV. After the ligation, there was no elevation of the PVP. An intraoperative liver biopsy showed a fatty liver (macrovesicular steatosis: 15%) without fibrosis.

Doppler ultrasonography done the following day showed the intrahepatic portal vein, which had not been detected preoperatively. There was no communication between the portal vein and the inferior vena cava (Fig. 5). The PVP remained below 15 mmHg until the catheter was removed 7 days after the operation. Laboratory data returned to normal within 2 weeks. The serum galactose was 0 mg/dl after drinking milk, and the patient's SpO_2 in room air and exertional dyspnea also improved. He was discharged 18 days after the operation without any complications.

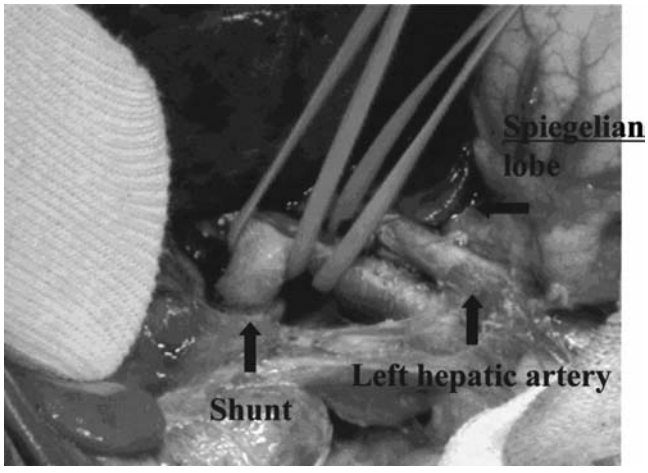


Fig. 3. The large portosystemic shunt between the portal vein trunk and the inferior vena cava

Discussion

Patent ductus venosus is rarely found in children, especially in association with hypergalactosemia and an intrapulmonary shunt.[2,3] This anomaly was first described by Barjon et al. in 1972.[4] The ductus venosus is a normal shunt in the fetus, which connects the umbilical vein to the inferior vena cava. Functional closure of the ductus venosus appears to be a consequence of a decrease in pressure in the portal sinus. With the postpartum closure of the umbilical vein, flow in the ductus venosus decreases and eventually ceases, with the final fibrotic transformation of the ductus venosus into the ligamentum venosum. The timing of this process is variable. The ductus venosus will remain open as a bypass tract of the portal vein after birth if there is increased vascular resistance in a poorly developed intrahepatic portal system, as in our patient.[5,6]

The term "hepatopulmonary syndrome" was proposed by Kennedy and Knudson[7] who suggested that it may be caused by a specific vasodilator unable to be metabolized because of liver failure or portal hypertension. Therefore, hepatopulmonary syndrome is very likely in a patient with a large portosystemic shunt.

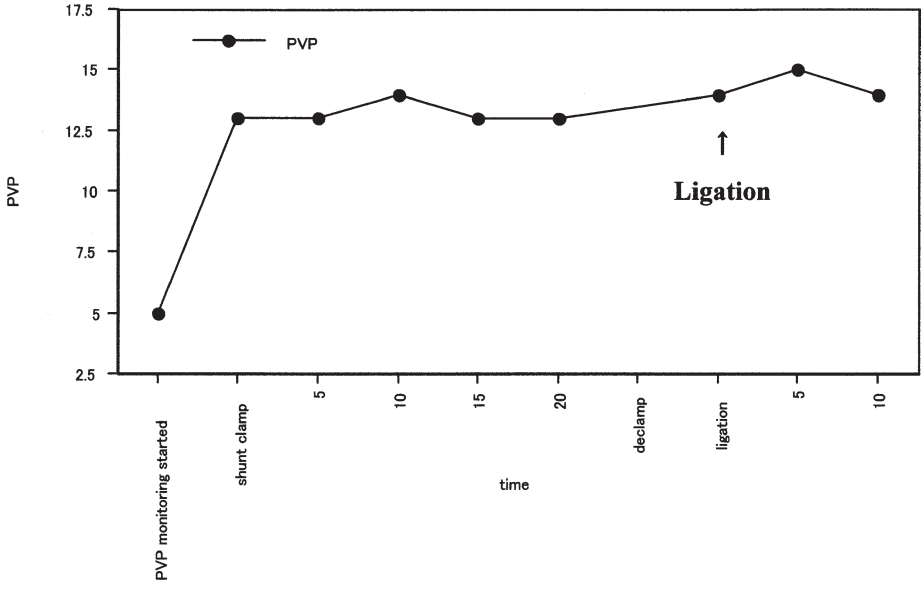


Fig. 4. Intraoperative portal vein pressure (*PVP*) monitoring. The portal vein pressure increased to within normal limits after clamping and ligation of the portosystemic shunt

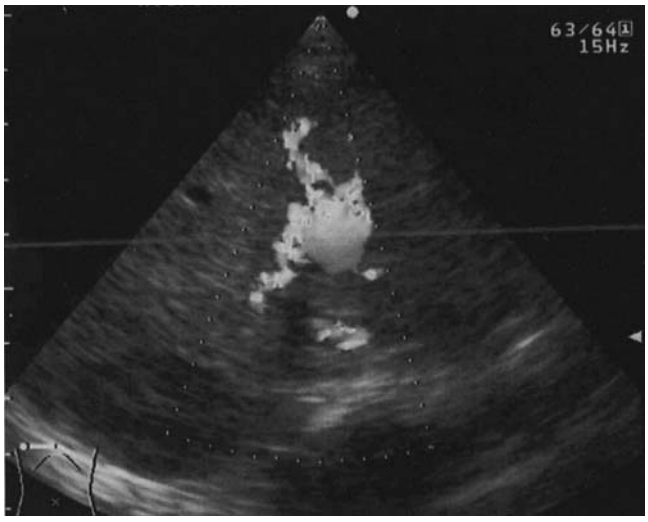


Fig. 5. Doppler ultrasonography done the day after the operation clearly showed the intrahepatic portal vein flow. There was no flow between the portal vein and the inferior vena cava

We considered that the surgical indications in our patient were hyperammonemia, hypergalactosemia,and the intrapulmonary shunt. Egawa et al.[8] reported that when liver transplantation was performed for patients with a moderate intrapulmonary shunt, the shunt ratio decreased and resolved within a few months. Some patients with a shunt size of less than 5 mm have been successfully treated by interventional embolization,[9,10] whereas others have been treated by constricting the PDV by surgical banding without complete occlusion.[11,12] If the PDV does not close eventually after banding, reoperation is required for complete ligation. Banding is the preferred approach if the PVP increases after test clamping of the PDV. Reoperation was needed for ligation in one patient and another patient with malformation of the hepatic vein went into shock 5 h after surgery, necessitating release of the band. Although liver transplantation could be chosen as a treatment for PDV, it is often followed by complications.[13] On the other hand, Barsky et al.[14] reported that ligation of the shunt may be followed by life-threatening complications, and stated that the portal venous system must be carefully assessed before attempting closure of a PDV. Therefore, we propose the following treatment selection for PDV according to severity.

1. For a small shunt without hypoplasia of intrahepatic portal vein: interventional embolization
2. For a large shunt without malconformation of the hepatic vein: ligation or banding under intraoperative PVP monitoring
3. For fibrosis confirmed by liver biopsy or an increase in the PVP after ligation or stricture: liver transplantation

To our knowledge, this is the first case report of a child with collapsed intrahepatic portal branches and an intrapulmonary shunt, in which excellent improvement of the intrahepatic portal vein flow was achieved by ligation of the large ductus venosus under intraoperative PVP monitoring. Thus, we suggest that intraoperative PVP monitoring is beneficial in the surgical decision-making process for PDV. Furthermore, ligation under PVP monitoring could be an appropriate and effective treatment even if the shunt is very large.

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