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Evaluation of the portal vein after duodenoduodenostomy for congenital duodenal stenosis associated with the preduodenal superior mesenteric vein, situs inversus, polysplenia, and malrotation

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Abstract A male infant weighting 2970 g with total situs inversus, polysplenia, malrotation, duodenal stenosis, and complex cardiac anomalies, was admitted to our hospital. At 4 days of age, he underwent surgery that revealed a blood vessel passing over the duodenum from the mesenterium to the porta hepatis. A loose overbridging duodenoduodenostomy was performed to prevent compression of the vessel. The cardiac anomalies were corrected, and he could eat unrestricted diets. At the age of 1 year and 3 months, a 3-dimensional computed tomographic scan demonstrated that the vessel on the duodenum was the superior mesenteric vein (SMV), and it formed the portal vein with the splenic vein at the porta hepatis. Further, the scan revealed no compression of the SMV at the anastomosis. Doppler ultrasonography revealed a normal portal blood flow of 118.6 mL/min. This report describes the junction between the SMV and the splenic vein in a patient who had the SMV passing over the duodenum from the mesenterium. Correctly, patients previously diagnosed with a preduodenal portal vein could have a preduodenal SMV. The loose overbridging duodenoduodenostomy had advantages not only in passage of the anastomosis but also in maintenance of the portal blood flow for the congenital duodenal obstruction with the preduodenal SMV.

Several procedures of anastomosis for congenital duodenal obstruction associated with a preduodenal portal vein (PDPV) have been reported [1-5]. However, there has been no study that investigated the postoperative portal blood flow in these cases. We investigated the connection of the SMV and the splenic vein (SV), and the patency and blood flow in the portal vein after performing a loose overbridging duodenoduodenostomy for a congenital duodenal stenosis associated with a blood vessel passing over the duodenum from the mesenterium.

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1. Case

A male infant weighing 2970 g was born at 40 weeks of gestation. At 27 weeks of gestation, he was discovered to have polyhydramnios, gastric dilatation, situs inversus, and cardiac anomalies. Using a cardiac ultrasonography in combination with a gastrointestinal contrast study, he was diagnosed as having total situs inversus, malrotation, duodenal stenosis, dextrocardia, double-outlet right ventricle, hypoplastic left heart syndrome, single ventricle, right aortic arch, and pulmonic stenosis after birth. At the age of 4 days, he underwent an abdominal operation that revealed the dilated stomach on the right side of the abdomen and the normally right-sided liver on the left. The several small spleens were found at the right extremity of the liver. There was a thick blood vessel passing over the first portion of the duodenum from the mesenterium to the porta hepatis, and the oral duodenum was dilated. The colon was lying on the second portion of the duodenum, which continued to the jejunum in the left abdomen. The colon was parted from the duodenum to the right abdomen. To clarify the cause of the stenosis, the duodenum was opened transversely on the oral side at approximately 10 mm from the vessel. Although there was an intrinsic membrane at the vessel, communication to the anal side could not be found. A duodenoduodenostomy overbridging the vessel was attempted. The duodenum was opened longitudinally on the anal side at approximately 10 mm from the vessel, and to avoid compression of the vessel, a loose side-to-side duodenoduodenostomy was performed. After the anastomosis, the blood flow in the vessel surrounded by the duodenum was efficiently maintained (Fig. 1). The patient underwent Blalock-Taussig's shunt operation at 1 month of age, Glenn's operation at 10 months, and Fontan's operation at 1 year and 3 months. The course following the surgeries was uneventful, and the patient has responded well. At 1 year and 3 months of age, after completion of a weaning diet, the patient was able to eat unrestricted diets without vomiting; his height was 75 cm and his weight was 8.2 kg (within normal). A gastrointestinal contrast study, an enhanced 3-dimensional computed tomography (3D-CT) scan, and Doppler ultrasonography were performed to estimate the passage through the duodenal anastomosis, the compression of the vessel at the anastomosis, and the portal blood flow. The gastrointestinal contrast study showed no dilatation of the oral duodenum and a clear passage. The 3D-CT scan established that the vessel at the duodenal anastomosis was the SMV; this vessel showed no evidence of compression. Furthermore, it revealed that the SMV and the SV formed the portal vein at the porta hepatis, and the portal vein branched into the 3 intrahepatic portal veins (Fig. 2). The color Doppler ultrasonography showed the hepatopetal blood flow, and pulse Doppler ultrasonography measured his portal blood flow as 118.6 mL/min.

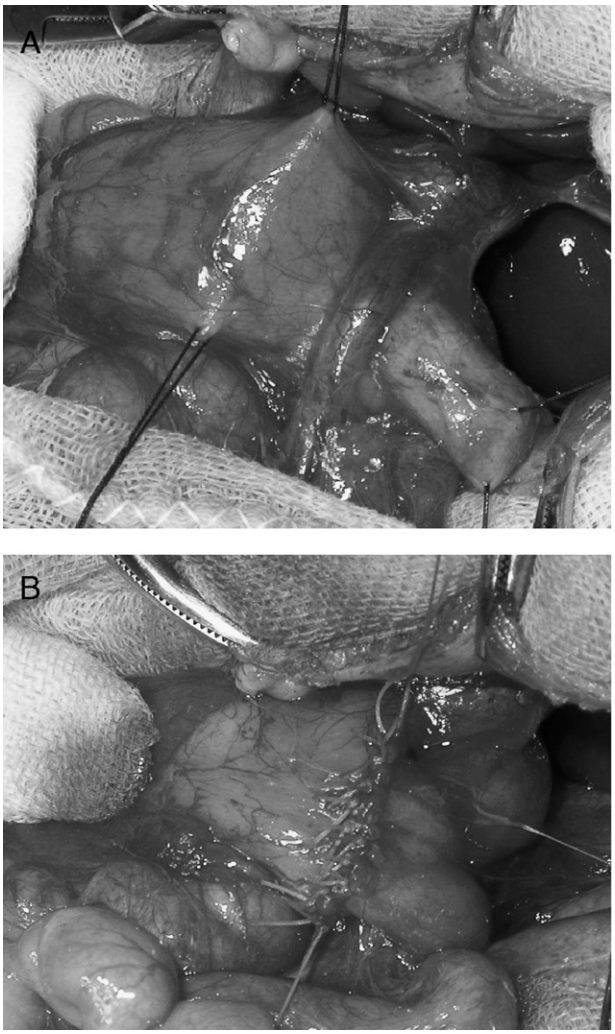

Fig. 1 An abdominal operation revealed that a blood vessel passed over the first portion of the duodenum from the mesenterium to the porta hepatis, and the oral duodenum was dilated (A). The duodenum was opened transversely on the oral side at approximately 10 mm from the vessel and longitudinally on the anal side at approximately 10 mm from the vessel. After a loose overbridging side-to-side duodenoduodenostomy was performed, the blood flow of the vessel surrounded by the duodenum was efficiently maintained (B).

2. Discussion

The spectrum of heterotaxia syndrome (HS) comprising situs inversus, polysplenia, and asplenia, is caused by the disorder of heart looping and intestinal rotation during the early weeks of gestation, and it has many variations [3]. More than 40% of patients with HS have serious congenital cardiac anomalies [3,6], and approximately 40% of patients suffer from congenital gastrointestinal anomalies. It has also been reported that HS patients associated with congenital vascular anomalies and that the PDPV is implicated in 17% to 50% of them [6,7]. Because congenital duodenal obstruction occurs in 50% to 80% of the patients with a PDPV, newborn babies