Thrombus formation within hepatic vein after Fontan procedure

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Thrombus formation is an important complication after Fontan procedure.¹ Although thrombus formation is commonly seen either in the systemic venous pathway or in the pulmonary venous pathway, to the best of our knowledge there have been no reports of thrombus formation within the hepatic vein. Here we present two cases of unusual intrahepatic thrombus formation after the Fontan procedure.

Clinical Summaries

PATIENT 1. A 4-year-old girl was being followed up at our hospital for the diagnosis of pulmonary atresia with intact ventricular septum. She had undergone a bidirectional cavopulmonary shunt at the age of 3 years. In September 2004, she underwent a Fontan procedure. The procedure consisted of anastomosis between the proximal stump of the superior vena cava (SVC) and the pulmonary artery and creation of lateral tunnel from the inferior vena cava (IVC) to the orifice of the SVC. A tunnel of sufficient caliber was created with in situ atrial free wall. Postoperative echocardiography showed good cardiac function and unobstructed cavopulmonary pathway. For anticoagulation, drip infusion of heparin and oral intake of aspirin were started on postoperative day (POD) 1. On POD 3, blood data showed elevation of hepatic enzymes (aspartate aminotransferase 1010 IU/L, alanine aminotransferase 940 IU/L). Abdominal echography showed dilated hepatic veins and a thrombus 10 mm in diameter at the confluence of right, middle, and left hepatic veins (Figure 1). Therapy with warfarin was started to prevent further enlargement of the thrombus. The values of the hepatic enzymes dropped gradually and were within reference range on POD 14.

PATIENT 2. A 2-year-old boy had the same diagnosis as patient 1. He had undergone bidirectional cavopulmonary shunt at the age of 1 year. Creation of the lateral tunnel from the IVC to the orifice of the SVC was performed as in patient 1, and the postoperative course was uneventful. Thrombus at the confluence of hepatic vein and IVC was found by abdominal echography performed on POD 14. The thrombus extended into the middle hepatic vein, and part of it protruded into the IVC (Figure 1). There was neither elevation of hepatic enzymes nor hepatic venous dilation, and therapy with warfarin was started.

In both cases, preoperative measurements of hematologic parameters, serum protein, renal and liver function parameters, and coagulation status were within reference ranges. Cardiopulmonary bypass was instituted with ascending aortic perfusion, and bicaval venous drainage was performed with L-shaped venous cannulas inserted directly into the SVC and IVC.

In both cases, thrombi had disappeared within 2 weeks after the initiation of warfarin therapy (Figure 1), and lung perfusion scintigraphy showed no sign of pulmonary embolism. Both patients have been followed up for 2 years and show no signs of thrombus.

Discussion

We report 2 rare cases of intrahepatic thrombus formation after the Fontan operation. Several predisposing risk factors for the development of thrombi have been proposed. Slow and non-pulsatile flow through the caval veins and low cardiac output are inherent in the physiology of all patients after the Fontan operation, regardless of the type of cavopulmonary connection. In addition, some reports suggest that an imbalance in coagulation proteins after the Fontan operation may predispose toward the development of thrombus.^{2,3} In our 2 cases, there were

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Figure 1. *Upper panels* show thrombi formed within hepatic veins in patients 1 (*left*) and 2 (*right*). As shown in *lower panels*, thrombi in both patients 1 (*left*) and 2 (*right*) had disappeared within 2 weeks. *RHV*, Right hepatic vein; *MHV*, middle hepatic vein; *LHV*, left hepatic vein; *TH*, thrombus; *IVC*, inferior vena cava.

neither obstruction of cavopulmonary pathway nor significant abnormalities in coagulation proteins. Relative to other cases, there was no significantly increased physiologic risk factor that would precipitate the formation of thrombus.

The other explanation for the formation of the intrahepatic thrombi is possible hepatic venous endothelial injury at the time of cannulation to the IVC. In general, a distance between the Eustachian valve and the upper margin of the opening of the hepatic vein is short, and the tip of the venous cannula may be inserted incidentally. In a study with transesophageal echocardiography, the incidence of the venous cannula being primarily placed in the hepatic vein was nearly 10%.⁴ Both the endothelial injury and the slow flow velocity within cavopulmonary tract thus may have contributed to the formation of the hepatic venous thrombi in these 2 cases.

It has been our institutional approach to avoid routine anticoagulation therapy with warfarin after a Fontan operation unless the

extracardiac conduit is used as a systemic venous pathway. Short-term warfarin therapy after a Fontan operation might have prevented the formation of intrahepatic thrombi in these cases, because both occurred within 2 weeks after the operation.

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