A female child with visceral heterotaxy (presumed left isomerism), interrupted inferior vena cava with azygos continuation to the superior vena cava, and double-outlet right ventricle underwent a total cavopulmonary anastomosis (end-to-side anastomosis of the superior vena cava to the right pulmonary artery) in infancy. Completion of Fontan circulation was undertaken at 3 years of age with construction of a tunnel from the hepatic veins to the pulmonary artery confluence.

By 6 years of age, the child had become increasingly cyanotic, and cardiac catheterization was performed to define the mechanism of the cyanosis. Figure 1 is a pulmonary artery angiogram that reveals moderate hypoplasia of the pulmonary artery confluence. Figure 2 is a selective angiogram performed in the hepatic vein tunnel, with hepatic venous flow directed predominantly toward the left lung. The left pulmonary artery angiogram was normal. A right pulmonary artery angiogram (Figure 3) was markedly abnormal, showing a diffusely granular appearance of the distal vasculature and early appearance of contrast in the right pulmonary veins, characteristic of the presence of pulmonary arteriovenous malformations. The development of arteriovenous malformations in the right lung allowed for intrapulmonary right-to-left shunting, accounting for the patient’s progressive cyanosis. Figure 4 is an azygos vein angiogram, performed after the
implantation of an intravascular stent into the hypoplastic pulmonary artery confluence. Note the presence of a duplicated inferior vena cava (the left inferior vena cava received the left renal vein).

This case illustrates the importance of hepatic venous return circulating through the pulmonary vasculature in inhibiting pulmonary angiogenesis. The “antiangiogenic” properties of hepatic venous blood are attributed to an as-yet uncharacterized hepatic factor. The left lung, which received hepatic venous return through the surgically created hepatic vein tunnel, was normal. However, the right lung, which was underperfused with hepatic venous effluent both by virtue of the geometry of the hepatic vein to pulmonary artery tunnel and the narrowing of the pulmonary artery confluence, developed arteriovenous malformations.

Figure 3. Selective angiogram in right pulmonary artery (RPA). Note early appearance of contrast in right pulmonary veins (RPV) draining into pulmonary venous atrium (PVA) and granular appearance of distal pulmonary vasculature (compare with normal appearance of left pulmonary vasculature in Figure 2).

Figure 4. Angiogram in azygos vein (Az) depicting interruption of inferior vena cava with azygos continuation to superior vena cava. Large black arrow denotes a duplicated (left) inferior vena cava. Double small black arrows depict the intravascular stent implanted into hypoplastic segment between right (RPA) and left (LPA) pulmonary arteries.
Development of Unilateral Pulmonary Arteriovenous Malformations due to Unequal Distribution of Hepatic Venous Flow
Henri Justino, Lee N. Benson and Robert M. Freedom

Circulation. 2001;103:e39-e40
doi: 10.1161/01.CIR.103.8.e39

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/103/8/e39

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/