*Case Reports*

# Regression of a Large Congenital Hepatic Arteriovenous Malformation

**Hala Mounir Agha, MD**
**Rania Zakaria, MD**
**Fatma Alzahraa Mostafa, MD**
**Hala Hamza, MD**

*Congenital hepatic arteriovenous malformations are rarely seen in association with persistent neonatal pulmonary hypertension. We report the case of a full-term female newborn who presented with heart failure and respiratory distress soon after birth. Echocardiographic investigation revealed severe persistent pulmonary hypertension and patent ductus arteriosus. Here we report spontaneous regression in size of both the feeder vessel and the vascular bed of the congenital hepatic arteriovenous malformation. We postulate that our conservative use of oral heart failure therapy, in the form of diuretic agents and captopril, decreased the congestion and diameter of the affected vessels.* ***(Tex Heart Inst J 2015;42(2):184-7)***

Congenital hepatic arteriovenous malformations (HAVMs) are rare anomalies that typically present in infancy, together with congestive heart failure, anemia, and hepatomegaly. Morbidity and mortality rates are high if the condition is not recognized and treated promptly.[1] We report the regression of a large congenital HAVM that presented with heart failure and pulmonary hypertension, in early infancy.

## Case Report

In January 2010, a female term infant, who had been delivered vaginally, presented at the age of 20 days with respiratory distress. Her body weight was 6.6 lb, and she had mild tachypnea and diaphoresis upon feeding (class II, Modified Ross Heart Failure Classification for Children). Tachycardia (heart rate, 160 beats/min) was evident without desaturation (oxygen saturation on room air, 95%). Her femoral pulse was easily felt. There was no detectable cardiomegaly (apex beat at the 4th intercostal space inside the midclavicular line), but there was an accentuated $S_2$ with an audible soft systolic murmur at the tricuspid area. There was mild hepatomegaly, and chest auscultation revealed normal expansion without audible crepitations. Transthoracic echocardiography showed a dilated right ventricle with moderate tricuspid regurgitation and an estimated systolic pulmonary artery pressure of 50 mmHg. There was a small patent foramen ovale with left-to-right flow and a tiny ductus arteriosus. No left atrial or left ventricular dilation was detected. At least 2 pulmonary veins were draining normally into the left atrium, but an abnormal subdiaphragmatic structure was noted within the liver, during sonographic examination. The patient did not have any skin or mucosal telangiectasis or hemangioma, and the family history of hereditary hemorrhagic telangiectasia was negative.

On the basis of the clinical and echocardiographic findings, the differential diagnosis included infradiaphragmatic total anomalous pulmonary venous drainage, congenital hepatic shunts, infantile hepatic hemangioma, and vascular malformations. Consequently, a multislice computed tomographic angiogram (CTA) of the heart and great vessels was ordered. Because the general condition of the baby was stable, oral heart failure therapy was prescribed in the form of furosemide (2 mg/kg/d), captopril (1.5 mg/kg/d), and digoxin (0.05 mg/kg/d). A 64-slice CTA obtained on a GE Lightspeed™ VCT (GE Healthcare; Waukesha, Wisc) revealed a large, undefined (but well-circumscribed) vascular structure, subdiaphragmatic and epigastric in location. This structure displayed uniformly opaque spaces. The left hepatic vein (markedly prominent), the right internal mammary artery, and the celiac trunk arterial branch were seen to be connected within this structure. Multiple enumerable smaller vessels

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***From:*** *Pediatric Department (Drs. Agha, Alzahraa Mostafa, and Hamza), Children Hospital, Cairo University, Cairo 11562; and Radiology Department (Dr. Zakaria), Cairo University Hospital, Cairo 11513; Egypt*

***Address for reprints:*** *Hala Mounir Agha, MD, Pediatric Cardiology Unit, Pediatric Department, Faculty of Medicine, Cairo University, Kasr Al Aini St., Cairo 11562, Egypt*

***E-mail:*** *halaazza@gmail.com*

were seen along the margins of this vascular structure and along the inferior surface of the diaphragm (Fig. 1).

During the follow-up period, extending to 4 months after hospital discharge, we noticed substantial clinical improvement of the infant's symptoms (to class I, Modified Ross Classification). Auscultation revealed normal intensity of the $S_2$ (no cardiomegaly) and no audible murmur at the tricuspid area. Serial Doppler echocardiograms revealed the complete regression of pulmonary hypertension. A multislice CTA showed a significant size reduction of the irregularly shaped subdiaphragmatic vascular lesion. The substantial hypertrophy of the left hepatic vein that drained this lesion was also markedly decreased. The HAVM displayed a heterogeneous parenchymal texture and irregular vascular channels that were still tortuous, but much smaller in size (Fig. 2). The patient's controlled heart failure (as indicated by objective values), decreased respiratory rate, and weight gain prompted us to discontinue the previously prescribed oral heart failure therapy when she reached 6 months of age.

## Discussion

Peripheral arteriovenous malformations (AVMs) in the newborn are rare anomalies that can be associated with persistent pulmonary hypertension of the newborn (PPHN) and can result in significant morbidity and mortality rates if not diagnosed early.[2] Infants with large peripheral AVMs might display pulmonary hypertension because of the substantial increase in pulmonary blood flow that is superimposed on the existing neonatal pulmonary venous drainage.[3] Although PPHN has been reported in neonates with cerebral AVM,[4,5] we found only 2 reports in the literature of congenital HAVM in association with PPHN.[6,7] Congenital hepatic arteriovenous malformation, while rare, has a 50% to 90% mortality rate, which emphasizes the importance of a high degree of suspicion in achieving early diagnosis and definitive treatment.[2,8,9] Therefore, echocardiographic evaluation of PPHN should include a careful evaluation of the liver, to rule out HAVM.[1]

Here, we report the case of a term neonate with congenital HAVM who presented with heart failure and persistent pulmonary hypertension. It is a highly unusual case, if only because there was regression of both the feeder vessel and the vascular bed of this congenital malformation. Our decision to proceed with oral medical therapy, rather than with the usual interventional embolization of the abnormal intrahepatic vascular channel, arose from the clinical status of the infant. The baby had mild heart failure (class II, Modified Ross Classification), was hemodynamically stable, and was not in need of hospitalization. Under close follow-up, the patient received therapy at home, showed good clinical response (class I Modified Ross Classification) to the medical therapy, and was thriving. In addition, echocardiograms obtained 1 and 2 weeks later revealed

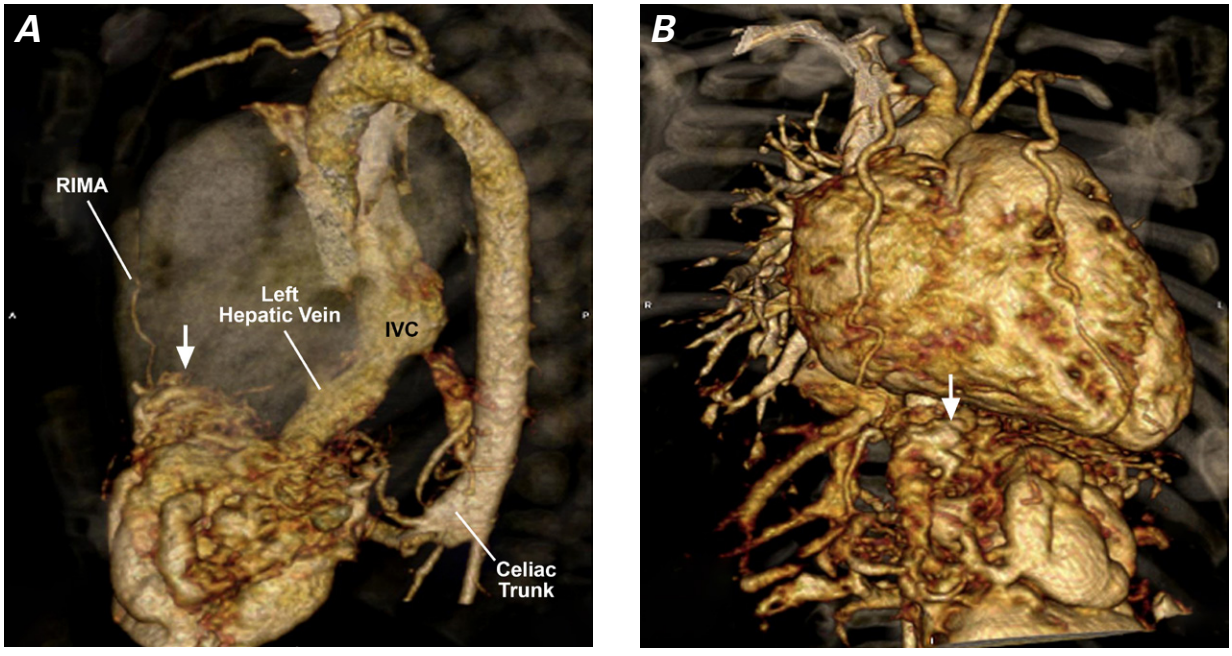


***Fig. 1*** *Computed tomograms (3-dimensional volume-rendered) in **A**) left lateral and **B**) anteroposterior views show a large, undefined (but well-circumscribed) vascular structure (arrows)—subdiaphragmatic and epigastric. The celiac trunk and right internal mammary artery (RIMA) supply this vascular structure. The left hepatic vein is prominent and appears to be draining the anomaly.*

*IVC = inferior vena cava*

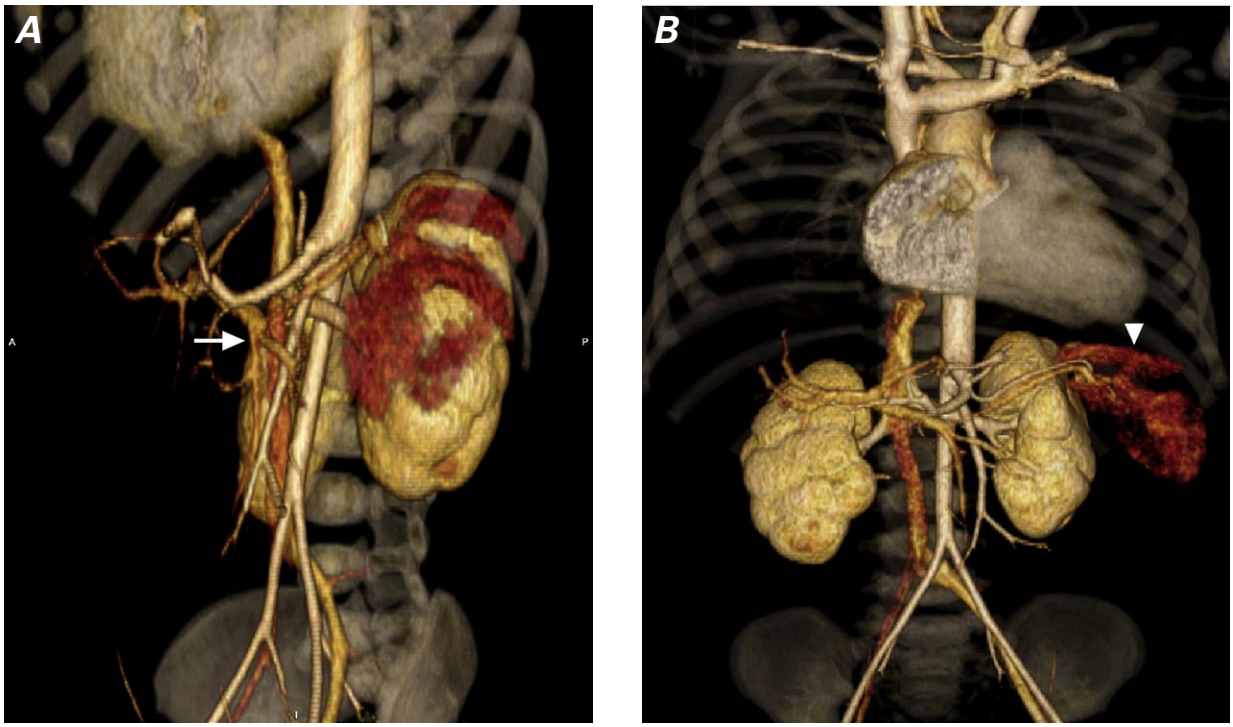


***Fig. 2*** *Four months later, computed tomograms (3-dimensional volume-rendered) in* ***A****) left lateral and* ***B****) anteroposterior views show that the left hepatic vein (arrow) draining the anomaly is still prominent, but that the vascular lesion itself (arrowhead) is markedly reduced in size.*

gradual reduction of systolic pulmonary artery pressure and right ventricular size.

These cumulative data encouraged us to continue our plan of conservative management. We postulate that using oral heart failure therapy, in the form of diuretic agents and captopril, decreased the congestion and diameter of the affected vessels. Captopril blocks the conversion of the inactive peptide angiotensin I to the active peptide angiotensin II, with a resulting decrease in peripheral vascular resistance,[10] a decrease in left-to-right shunting,[11] and a consequent chance for spontaneous thrombosis of this abnormal vascular channel.

Only one previous published article, which presented the cases of 4 neonates with AVMs (cerebral in one, hepatic in 2, and pulmonary in one), reported that neonates with hepatic malformations improved upon medical therapy. Those authors concluded that color-flow Doppler ultrasonography should be the gold standard for the diagnosis of cerebral and hepatic malformations, and that selective angiography should be reserved for patients who might benefit from embolization or surgical resection.[12]

The case reports of congenital HAVM are scarce, as a consequence of the rarity of the malformation itself. However, the unexpected good outcome for our patient can be interpreted by revisiting the natural history of the vein of Galen. The suggested causes of spontaneous thrombosis of aneurysmal malformation in the vein of Galen include slow-flow shunts, obstruction of the venous outflow tract, and obstruction of the feeding artery.[13,14] We postulate that, in our patient, medical therapy in the form of diuretic agents and captopril decreased the diameter of the left-to-right shunt and the hepatic vein diameter and enabled venous thrombosis of that vascular channel.

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