Congenital Arteriovenous Malformations Between Brachiocephalic Arteries and Systemic Veins*

Fernando R. Gutierrez, M.D., F.C.C.P.;‡ Michele P. Monaco, M.D.;§ Alexis F. Hartmann, Jr., M.D.;‡ and Robert C. McKnight, M.D.†

Three patients under two years of age are described with unusual fistulas involving the brachiocephalic arteries and the innominate vein or the superior vena cava. Two patients were asymptomatic, and one newborn was cyanotic and in congestive failure. This unusual presentation has only rarely been reported in thoracic arteriovenous malformations.

Arteriovenous fistulas may be congenital or acquired. Congenital arteriovenous fistulas are uncommon anomalies which have been reported primarily in the brain, neck, thoracic wall, heart, liver, and extremities.¹ ² We describe three patients with arteriovenous malformations involving branches of the brachiocephalic vessels and drainage into the superior vena cava system.

CASE REPORTS

CASE 1

This 2,900-gram infant was delivered by cesarean section and was noted to be cyanotic and in respiratory distress. Arterial oxygen on room air was 25 mm Hg and 120 mm Hg on 100 percent oxygen via hood. A continuous loud murmur accompanied by a thrill was heard over the left and right sternal borders at the 2nd and 3rd intercostal spaces, with radiation to the neck, occipital region, and also to the left antecubital fossa. A chest roentgenogram showed generalized cardiomegaly with mediastinal enlargement in the area of the superior vena cava (Fig 1). The ECG suggested right atrial enlargement. A contrast M-mode echocardiogram revealed a right-to-left atrial level shunt appearing four cardiac cycles after an IV injection. A cranial sonogram was within normal limits. Cardiac catheterization revealed oxygen saturations of 81 percent in the superior vena cava, 81 percent in the right atrium, and 76 percent in the inferior vena cava. Angiography showed a left subclavian artery-to-innominate vein fistula that drained into a markedly dilated superior vena cava (Fig 2a and b). A median sternotomy was performed and the fistula ligated. Despite a complicated postoperative course, follow-up at ten months of age revealed the infant to be asymptomatic with no cardiac murmur. Recatheterization showed no residual fistula.

CASE 2

A 19-month-old girl was found to have a grade 2–3/6 continuous

murmur at the right and left upper sternal borders at the 2nd and 3rd intercostal spaces, not affected by pressure on the neck veins. The pulses were bounding. The ECG was suggestive of biventricular enlargement. Her growth and development were normal. The child was studied by catheterization and angiography and was shown to have an arteriovenous malformation between the right subclavian artery and the azygous vein system (Fig 3). The child was operated on and the fistulous arterial supply ligated. She made an uneventful recovery.

CASE 3

A three-month-old boy, completely asymptomatic, was found to have a continuous murmur along the right and left sternal borders at the 2nd and 3rd intercostal spaces. An ECG and chest roentgenogram were normal. A Doppler flow study suggested a left-
right aortic level shunt. Cardiac catheterization and angiography revealed an extensive arteriovenous malformation involving the left cervical paravertebral area, originating from an anomalous arterial branch from the aortic arch, draining to the superior vena cava (Fig 4). Because of its extensive nature, the lack of symptoms, and the possibility of spinal vascular supply involvement, the child has been followed without surgical intervention. He is currently 7½ years old and continues to have a grade 2/6 continuous murmur at the left base, with wide radiation diffusely over the upper thorax, but is otherwise without symptoms.

**DISCUSSION**

Arteriovenous malformations involving the thoracic cavity are uncommon. Most reported cases may be classified as acquired and are secondary to trauma. The congenital forms are even more uncommon, and because of their extremely varied clinical presentations are frequently a diagnostic challenge. We report three cases of the congenital form demonstrating this wide spectrum of clinical and anatomic findings.

Asymptomatic or mildly symptomatic thoracic arteriovenous fistulas have been reported involving brachiocephalic, intercostal, and internal mammary arteries. Our cases 2 and 3 exhibited a similar lack of severe symptoms; however, the size and complexity of the malformation is sometimes surprising (Fig 4).
The majority of arteriovenous fistulas that produce high-output congestive heart failure will do so in the neonatal period and usually involve the head (vein of Galen), liver (hepatic hemangioendothelioma), or kidneys. Only two other cases have been reported involving the thorax. Progressive cyanosis has been ascribed to the presence of congestive failure and edema in the lungs or to intrapulmonary shunting. Evidence for persistence of the fetal circulation with a right to left shunting at the atrial or ductal level (see "Patient 1") may further increase the cyanosis or explain the presence of cyanosis in infants with clear lung fields.

The most important physical findings in all the reported cases of thoracic arteriovenous fistulas are a continuous murmur with a wide pattern of radiation and an occasional accompanying thrill. Pulses may be bounding dependent on the size of the malformation. An increase in oxygen saturation in the superior vena cava or its branches may be helpful in the diagnosis (case 1), and echocardiography may be of considerable help in suggesting the diagnosis as well as excluding cardiac causes of congestive failure or cyanosis. An-