Heart Failure and Cyanosis in a Newborn*

William E. Hellenbrand, M.D.;** Michael J. Kelley, M.D.; and Michael A. Berman, M.D.

A 3.2 kg girl was delivered following a labor complicated by fetal distress. Apgar scores were 3 and 5. At 12 hours of age, her respiratory rate was 80/min, with a heart rate of 170/min. The pulse in each extremity was markedly diminished and the blood pressure was 50/30 mm Hg. There was a grade 4/6 holosystolic murmur at the left lower sternal border radiating to the left axilla and to the right sternal border. No murmur was audible over the head or the liver. The liver was enlarged 6 cm below the right costal margin. The Po2 was 35 mm Hg in room air and 55 mm Hg on 100 percent O2 An electrocardiogram showed combined ventricular and atrial enlargement and significant ST-T wave changes. The echocardiogram demonstrated two dilated ventricles, a large left atrium, and four normal cardiac valves. The chest roentgenogram is presented in Figure 1.

*From the Departments of Pediatrics and Diagnostic Radiology, Yale University School of Medicine and Yale-New Haven Hospital, New Haven.
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Reprint requests: Dr. Hellenbrand, 333 Cedar Street, New Haven 06510

FIGURE 1
Diagnosis: Cerebral arteriovenous malformation involving the vein of Galen

The chest roentgenogram (Fig 1) shows marked cardiomegaly with increased vascularity in the lungs. At cardiac catheterization, intracardiac pressures were consistent with right and left heart failure. Saturation data demonstrated a large oxygen step-up on the right side of the heart localized to the internal jugular vein. Right-to-left shunting at the pulmonary and foramen ovale levels was present. A transverse thoracic aortogram revealed a large arteriovenous fistula draining into the vein of Galen as shown (Fig 2). The AP and lateral views of the head demonstrate the arterial phase of the cerebral angiogram. The left vertebral artery (VA) and left internal carotid artery (LICA) and their branches are visualized. The basilar artery (BA) and the posterior cerebral artery (PCA) give rise to multiple tortuous arterial branches which empty into a markedly dilated vein of Galen (large white arrows). The lack of good arterial filling of the anterior (A) and posterior (P) portions of the brain indicates significant arterial steal due to the large arteriovenous malformation. Isoproterenol (Isuprel) was infused prior to surgery and a systolic bruit was now heard over the anterior fontanelle. The patient died during the operative attempt to ligate the multiple arterial vessels supplying the malformation.

An arteriovenous malformation involving the vein of Galen is a rare anomaly in which a variable-sized arteriovenous shunt exists between the cerebral circulation (invariably the posterior circulation) and the vein of Galen or venous channels proximal to it. Embryologically, the initial lesion is thought to be a congenital fistula between the choroidal vessels and the great cerebral vein. Such communications cause aneurysmal dilatation of the vein of Galen presumably because the vein is the most proximal distensible venous structure draining the shunts.1

In a neonate with congestive heart failure, cyanosis, decreased pulses, and a chest film demonstrating cardiomegaly and increased pulmonary vascularity, a systemic arteriovenous malformation should be considered. The two most common sites are the liver and the brain. This malformation may be responsible for intrauterine fetal stress and may cause congestive heart failure before or soon after birth. The large low resistance runoff through the fistula competes in utero with the placenta, the other low resistance circuit. High output failure in the fetus results from the large shunt through the malformation, producing a volume load on the heart.2 Other lesions that cause intrauterine failure (tricuspid insufficiency and absent pulmonary valve) also produce an enlarged heart. Normal or decreased pulmonary vascularity on the chest roentgenogram allows differentiation. The murmur in our patient resulted from dilatation of the cardiac chambers and secondary insufficiency of the atrioventricular valves. The absence of the cranial bruit prior to isoproterenol infusion was related to the severe congestive heart failure.

Most patients with a peripheral arteriovenous malformation demonstrate bounding arterial pulses. The systolic pressure is increased in response to the elevated cardiac output and the diastolic pressure is lowered secondary to the decreased peripheral resistance. It has been shown in experimental arteriovenous fistulas that flow and pressure distal to the lesion are decreased secondary to the runoff of blood into the fistula.3 This phenomenon, in addition to
congestive heart failure, explains the observed diminution in pulse pressure in the present case. Following initiation of isoproterenol (and presumably increased cardiac output), the carotid pulsations were prominent, but the peripheral pulsations remained decreased.

Persistence of fetal pathways and intrapulmonary shunts have been documented in patients with arteriovenous malformation. In the present case, the right-to-left shunt was at the atrial and pulmonary levels and explained the failure of the arterial Po2 to rise with increased ambient oxygen.

Thus, in a newborn with perinatal congestive heart failure, cyanosis, increased pulmonary vascularity, and cardiomegaly on the chest film, arteriovenous fistula must be considered even in the presence of decreased pulses. Careful palpation of the carotid pulses may be helpful. Rapid evaluation by cardiac catheterization and angiography may allow an opportunity for the fistula to be ligated.

REFERENCES
2 Talner NS, Campbell ACM: Recognition and management of cardiologic problems in the newborn infant. Prog Cardiovasc Dis 15:159-189, 1972