The patient was a 14-year-old girl admitted for evaluation of recurrent congestive heart failure. She had limited exercise tolerance and, although able to participate in sports, was fatigued easily. She also reported that her right leg was longer than her left and that her right shoe size was one size larger than the left. On physical examination, the above findings were confirmed, and an increased girth to the right thigh was noted as well. The admission frontal roentgenogram (Fig 1) shows a right-sided aortic arch and increased vascularity, while the lateral view (Fig 2) demonstrates some focal encroachment on the posterior tracheal wall.
Diagnosis: High cardiac output state secondary to a large arteriovenous malformation of the right thigh; right-sided aortic arch.

Cardiac catheterization confirmed the right-sided aortic arch but demonstrated no intracardiac anomaly. The resting cardiac output was 15.7 L/min, increasing to 27.2 L/min with exercise.

An arteriogram of the pelvis and right thigh demonstrated marked hypertrophy of the arterial tree of the right pelvis and right thigh which fed a large arteriovenous malformation, extending approximately from the pelvis to the distal part of the right femur (Fig 3). Feeders to this arteriovenous malformation were seen as high as a hypertrophied right lumbar artery at the L-4 level down to enlarged geniculate vessels from the popliteal artery.

In this case, the right-sided aortic arch was associated with an aberrant left subclavian artery as evidenced by the soft tissue opacity encroaching on the posterior wall. There are two major types of right-sided aortic arch. In the first type, known as mirror-image branching, there is no retroesophageal component to displace the esophagus and trachea anteriorly. Thus, the tracheal shadow appears normal on the lateral chest roentgenogram. This type of right aortic arch is associated with congenital heart disease in 98 percent of cases. In the other type of right aortic arch, that with an aberrant left subclavian artery, the left subclavian artery arises from a diverticulum which is posterior to the esophagus. With this type of right arch, there is almost always a soft tissue shadow encroaching on the posterior wall of the trachea on the lateral roentgenogram. With such an arrangement, there is only a 12 percent incidence of congenital heart disease.1,2

Our patient's increased vascularity and recurrent episodes of congestive heart failure were due to her massive arteriovenous malformation. The increased cardiac output in such patients is necessary to maintain blood pressure in the face of decreased systemic vascular resistance. In compensation for such decreased resistance, the heart rate and stroke volume increase, the diastolic blood pressure falls, and the cardiac output rises.3

The ability of systemic arteriovenous fistulas to cause increased cardiac output and high output congestive heart failure is well known. Cerebral and hepatic fistulas are probably the most common, although limb, pelvic, internal mammary, and epigastric vessels have also been reported.4,5 Most limb arteriovenous fistulas occur in the lower extremity.6 A systemic arteriovenous malformation should always be considered in a young patient with a large cardiac output and congestive heart failure, especially if no intracardiac defects are identified.

REFERENCES
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