SECTION EDITOR: WALTER W. TUNNESSEN, JR, MD

Picture of the Month

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A 6-WEEK-OLD INFANT, the product of a full-term, uncomplicated pregnancy and the first child of unrelated parents, had a history of irritability and poor feeding. The mother reported that the infant had been irritable and crying for a few days, especially during feeding, with episodes of pallor and breathlessness.

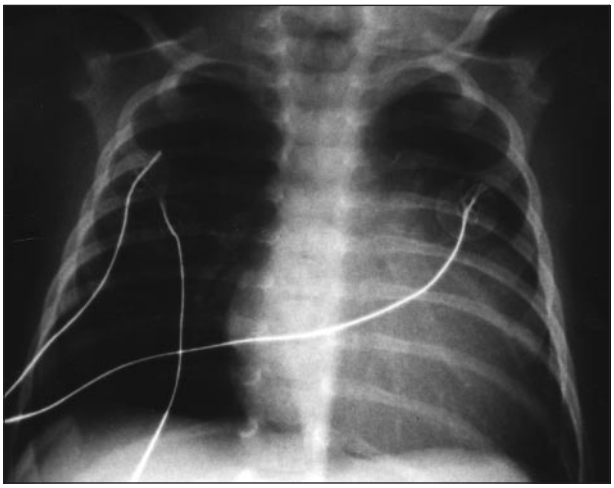

Figure 1.

On physical examination, the infant appeared well nourished and had no dysmorphic features. She was irritable and cried constantly. Her respirations were 50/min with intermittent grunting; pulse, 160/min; and temperature, 37°C. Oxygen saturation was 97% by pulse oximetry. Her skin was pale and extremities cool. On chest examination there were mild intercostal retractions, clear lung fields to auscultation, and a grade 3/6 systolic murmur along the left sternal border with a loud second heart sound and a gallop rhythm. The abdomen was soft with the liver edge palpable 4 cm below the right costal margin. Pulse and blood pressure were equal in all 4 extremities. A chest x-ray film (**Figure 1**) and electrocardiogram (**Figure 2**) were obtained.

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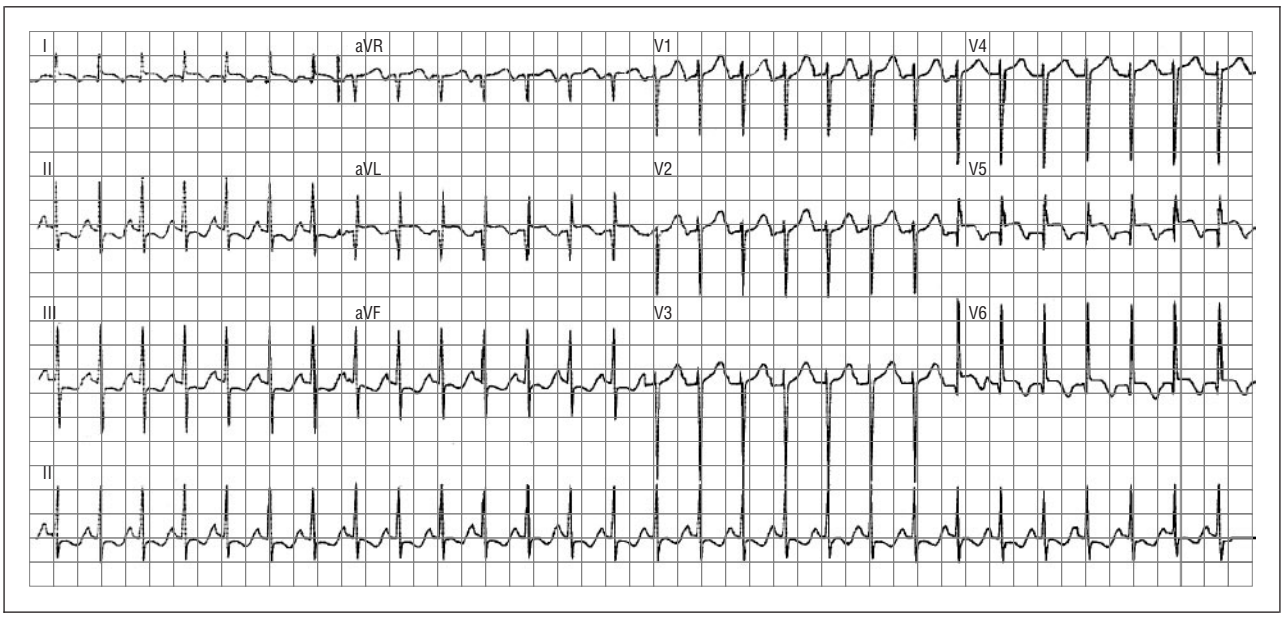

Figure 2.

Denouement and Discussion

Anomalous Origin of the Left Coronary Artery

Figure 1. *Cardiomegaly with pulmonary venous congestion is seen on the chest x-ray film.*

Figure 2. *The electrocardiogram shows deep Q waves with ST segment elevation and T-wave inversion in leads I, aVL, V_5, and V_6.*

The clinical presentation coupled with evidence of cardiomegaly and congestive heart failure on chest x-ray film and the electrocardiogram features of anterolateral myocardial infarction suggested the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery, which was subsequently confirmed by angiocardiography.

EMBRYOLOGY AND PATHOPHYSIOLOGY

The embryologic origin of the anomalous left coronary artery is either an abnormality in the septation of the conotruncus into the aorta and pulmonary artery or an abnormal persistence of the pulmonary buds and involution of the aortic buds that form the coronary arteries.[1]

Anatomically in this anomaly, the left coronary artery arises from the pulmonary artery, while the right coronary artery arises normally from the anterior aortic sinus. During fetal life, perfusion of the myocardium via the pulmonary artery is at pressures and oxygen saturation similar to those of the aorta. After birth, pulmonary artery pressure falls, and perfusion of the left coronary artery is compromised. In addition, pulmonary arterial blood is relatively desaturated, creating an inability to meet the high oxygen demand of the left ventricular wall, leading to myocardial ischemia. In response to these changes, the anomalous coronary artery dilates, and collateral vessels from the normal right coronary artery may develop. Since pressure in the anomalous left coronary artery is lower than in the right, blood may preferentially flow from the right coronary artery through collateral vessels into the left coronary artery and retrograde into the pulmonary artery, creating a coronary steal.

The degree of collateral flow and resultant perfusion of the anterolateral wall of the left ventricle determine when and if myocardial ischemia will occur. If collateral circulation is poor, symptoms and signs of myocardial ischemia occur early in infancy; but if well developed, individuals may reach adulthood without symptoms of myocardial muscle compromise.

CLINICAL FEATURES

In most of these infants, symptoms of myocardial ischemia, congestive heart failure, or myocardial infarction appear between 2 weeks and 6 months after birth, and include recurrent episodes of restlessness, irritability, incessant crying, and dyspnea, often associated with pallor and sweating. These episodes are most frequent during feeding, which corresponds to physical exertion for the infant. Irritability and crying are attributed to angina pectoris. Signs of congestive heart failure may be present, including tachypnea, tachycardia, gallop rhythm, cardiomegaly, and hepatomegaly. A murmur of mitral insufficiency may be heard, the result of infarction of a papillary muscle.

DIAGNOSTIC FEATURES

The electrocardiogram characteristically demonstrates an anterolateral infarction pattern with broad, deep Q waves in leads I, aVL, and the left precordium. These changes are often associated with persistent ST-segment elevation and T-wave inversion. Chest radiographs typically show moderate to severe cardiomegaly and pulmonary venous congestion. Echocardiography with Doppler color-flow mapping may successfully identify the anomalous vessels, but cardiac catheterization and angiography may be needed to demonstrate the anomalous anatomy and retrograde filling of the left coronary artery via collaterals from a dilated right coronary artery, with subsequent flow of contrast into the pulmonary artery.

PROGNOSIS

More than 80% of infants with this anomaly develop symptoms and signs of cardiac damage or failure in infancy, and about 65% to 85% die before 1 year of age.[2-4] Infants who survive generally have well-established coronary artery collateral circulation. These individuals may present later in childhood, adolescence, or even in adulthood with angina on effort or with congestive heart failure from mitral incompetence.[5,6]

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