Right Pulmonary Artery to Left Atrium Communication

An Unusual Cause of Cyanosis in the Newborn

John P. Cheatham, David A. Barnhart, Howard P. Gutgesell

The Lillie Frank Abercrombie Section of Cardiology, Department of Pediatrics, Baylor College of Medicine, and Texas Children's Hospital, Houston, Texas

SUMMARY. A one-day-old newborn infant presented with intense cyanosis, a continuous murmur, and mild congestive heart failure. The chest roentgenogram showed an abnormal right-heart border, and the echocardiogram demonstrated enlargement of the left ventricle and left atrium. Cardiac catheterization and angiography demonstrated a right pulmonary artery to left atrium communication. The infant responded favorably to medical management and is asymptomatic with the exception of mild cyanosis with crying. Right pulmonary artery to left atrium communication is a rare but potentially correctable cause of cyanosis in the newborn.

KEY WORDS: Right pulmonary artery to left atrium communication — Newborn cyanosis

Direct communication between the right pulmonary artery and the left atrium is a rare congenital cardiac defect. Cyanosis often accompanies this defect but usually is not evident until later childhood or adulthood. Described here is a newborn who presented with cyanosis at birth secondary to this anomalous communication, but whose condition spontaneously has improved.

Case Report

A 3.58-kg boy was born to a 20-year-old gravida 2, para 1 woman. Irregular fetal heartbeats were noted during labor, and a tight nuchal cord was present at delivery. The Apgar score was 1 at 1 minute. Immediate cardiopulmonary resuscitation was begun, and the Apgar score was 7 at 5 minutes. The baby continued to exhibit cyanosis and irregular heartbeats. The infant was transferred to Texas Children's Hospital, Houston, at 6 hours of age.

At the time of admission, examination revealed a cyanotic infant with a respiratory rate of 70 breaths per minute, pulse rate of 120 beats per minute and irregular, and blood pressure of 75/45 mm Hg. The lungs were clear to auscultation. The apical impulse was diffuse and displaced laterally into the left anterior axillary line. The S1 was normal and S2 was narrowly split with a normal pulmonary component. There was a prominent S3 at the apex. A grade 3/6 continuous murmur was heard at the upper left sternal border and was audible in the right hemithorax. The pulses were normal. The liver tip was palpable 3 cm below the right costal margin. Findings from the remainder of the physical examination were normal.

Laboratory Findings

The chest roentgenogram revealed cardiomegaly with normal pulmonary vascular markings. The contour of the right lower cardiac border was unusual, as seen in Fig. 1A. The ECG revealed extreme right-axis deviation, right ventricular predominance, generalized ST-segment changes suggestive of ischemia, and frequent premature ventricular contractions. An M-mode echocardiogram revealed a slightly enlarged left atrium (15 mm) and left ventricle (22 mm). The upper limits of normal for these dimensions are 14 mm and 20 mm, respectively [2]. The hemoglobin level was 17.4 g/dL, and the hematocrit reading was 52%. The WBC count was 16,000/cu mm, platelet count was 353,000/cu mm, calcium level was 10.4 mg/dL, and glucose level was 71 mg/dL. The arterial blood gas values in room air were PO2 of 30 torr, PCO2 of 39 torr, pH of 7.32, and an oxygen saturation of 51%. With administration of 40% oxygen by hood, the PO2 was 33 torr and oxygen saturation was 58%.
Fig. 1. The chest roentgenogram (A) demonstrates cardiomegaly. Note the unusual contour of the right cardiac silhouette as demonstrated by the arrows. Following injection of angiographic dye in the right pulmonary artery (B), the anomalous communication between the right pulmonary artery (RPA) and the left atrium (LA) is visualized (arrow). The aneurysmal dilatation of the left atrium corresponds to the abnormal cardiac silhouette as seen in Fig. 1A.

Hospital Course

Cardiac catheterization and angiography revealed a direct communication between the lower lobe branch of the right pulmonary artery and the left atrium (Fig. 1B). A patent ductus arteriosus and patent foramen ovale were also present. The right ventricular systolic pressure was 35 mm Hg, with no gradient across the pulmonary valve. The left ventricular systolic pressure was 65 mm Hg. The oxygen saturation was 85% in the left upper pulmonary vein, 66% in the left atrium, and 65% in the umbilical artery.

The baby was given digoxin, with rapid disappearance of the symptoms of congestive heart failure. The frequent premature ventricular contractions gradually diminished in number and were no longer present at the time of discharge at 11 days of age. The patient was asymptomatic at discharge, although he remained cyanotic.

Follow-up

On examination at four months of age, the infant was acyanotic at rest and had mild cyanosis with crying. Both respiratory and pulse rates were normal. There was a nonspecific grade 2/6 systolic murmur over the left sternal border. No continuous murmur was audible. The chest roentgenogram continued to show moderate cardiomegaly, and the ECG revealed right ventricular hypertrophy. Peripheral venous contrast echocardiography demonstrated evidence of intrapulmonary right-to-left shunting [3]. The M-mode echocardiogram (Fig. 2A) revealed contrast echoes appearing initially in the right ventricular outflow tract. After a delay of approximately 400 msec, the echoes appeared in the left atrium, indicating an extracardiac right-to-left shunt. The two-dimensional echocardiogram (Fig. 2B and C) allowed visualization of the microbubbles initially appearing in the right side of the heart without significant intracardiac right-to-left shunting. The microbubbles subsequently entered the left atrium through the anomalous communication from the pulmonary artery (Fig. 2D).

Our patient is asymptomatic at present, and surgical repair is planned when he weighs approximately 10 kg.

Discussion

Only 19 cases of direct communication between the right pulmonary artery and the left atrium have been reported in the literature as reviewed by Hiroshi et al [4]. Patients with this anomalous communication usually present with cyanosis, clubbing, and polycythemia. Sixteen of the 19 patients presented after 3 years of age. The oldest described patient was 45 years old at the time of diagnosis. The mean age at the time of presentation was 14 years.

Right pulmonary artery to left atrium communication is a rare cause of cyanosis in the newborn. Primary pulmonary disease or an intracardiac defect is the most common cause of cyanosis at this age. In our patient, normal pulmonary parenchymal markings on the chest roentgenogram and clear lung fields by auscultation made pulmonary disease an unlikely cause of cyanosis. The abnormal right cardiac silhouette on the chest roentgenogram seems to be a rather subtle but constant finding in patients with pulmonary artery to left atrium communication [5]. The continuous murmur that was well heard in the right axilla and over the right lower