A 5-month-old female baby presented to a community hospital in acute respiratory failure for which she was intubated and treated for presumed pneumonia. Chest x-ray demonstrated cardiomegaly, and a subsequent echocardiogram described mitral valve stenosis, mitral valve regurgitation, bicuspid aortic valve, perimembranous ventricular septal defect (VSD), and subaortic membrane, leading to the diagnosis of Shone complex. After several weeks of antibiotics, diuretics, mechanical ventilation, and inotropic support failed to improve her clinical course, the patient was transferred. Repeat echocardiography confirmed the previous anatomic findings, as well as moderately depressed left ventricular systolic function. Chalk sticking of the left ventricular papillary muscles was suggestive of an ischemic insult. Because of these findings, anomalous left coronary artery arising from the right pulmonary artery (ALCAPA) was suspected, but the left coronary artery (LCA) was not seen arising from the pulmonary root. A modified parasternal short-axis view revealed that the LCA arose anomalously from the mid right pulmonary artery (RPA; Figure 1). Flow within the anomalous LCA was bidirectional, although primarily antegrade, from the RPA toward the left ventricle (Movie I in the online-only Data Supplement). Cardiac catheterization confirmed this unusual diagnosis (Figure 2). The right coronary artery arose from the normal aortic position (Figure 3). The Qp:Qs ratio was 1.8 and indexed pulmonary vascular resistance was 3.2 Wood units m⁻². No significant mitral valve stenosis was found. Gradient through the VSD was 18 mmHg, and RPA saturation was 77% compared with a superior vena cava saturation of 63%. Interestingly, ECG obtained at the time of admission showed evidence of biventricular hypertrophy but no evidence of myocardial ischemia or infarction (Figure 4). The patient was taken to the operating room the following day, and the LCA was resected on a button of RPA tissue and reimplanted into the posterior aspect of the proximal aorta. Primary repair of the VSD was performed. She was discharged home on post-operative day 16. Predischarge echocardiogram demonstrated some improvement in left ventricular function.

ALCAPA was first described anatomically in 1865 by Krause and in 1885 by Brooks.1 ALCAPA accounts for <0.5% of congenital heart disease and is thought to occur in ≈ 1 in 300000 births.2 Typically, the LCA arises directly from the main pulmonary trunk adjacent to and in close proximity to the normal aortic origin as an isolated defect. However, rare
associations with VSD, patent ductus arteriosus, tetralogy of Fallot, or coarctation of the aorta have been described. More than 87% of ALCAPA cases present during infancy at ≈2 months of age, when pulmonary vascular resistance nadirs and preferential flow away from the myocardium into the lower-resistance pulmonary circuit create a coronary steal phenomenon, resulting in severe left ventricular and mitral valve dysfunction. Affected patients often present with

Figure 2. Angiography with an angiographic catheter positioned in the distal right pulmonary artery (RPA) with a balloon inflated to fill the proximal RPA and to prevent distal antegrade flow, demonstrating antegrade flow through the left coronary artery (LCA) and its branches in both the anteroposterior (A) and lateral (B) views.

Figure 3. Aortic root angiography demonstrating antegrade flow into the normally positioned right coronary artery (RCA) with an absence of filling of the left coronary artery as seen from the anteroposterior (A) and lateral (B) views. Ao indicates aorta.

Figure 4. A 12-lead ECG. Note the lack of deep q waves in leads I, aVL, and V₄ through V₆ classically seen with anterolateral myocardial infarct secondary to anomalous left coronary artery arising from the right pulmonary artery.
nonspecific symptoms such as poor feeding, respiratory distress, and fussiness that may obfuscate the diagnosis of congestive heart failure. A majority of untreated patients with ALCAPA die of congestive heart failure within the first year of life. Those who survive infancy may be at high risk of sudden cardiac death. Classically, the resting ECG demonstrates signs of anterolateral ischemia with deep q waves noted in leads I and aVL.

The presence of the VSD in our patient may have blunted these effects by allowing a degree of pulmonary hypertension and overcirculation, resulting in increased pressure through the RPA and increased oxygen content of blood entering the right coronary artery. Furthermore, the typical drop in pulmonary vascular resistance within the first month of life was delayed in this patient. We hypothesize that the combination of VSD-related mild elevation of RPA pressure and increased oxygen saturation, along with continued elevation in pulmonary vascular resistance, likely allowed antegrade flow through the anomalous LCA to the left ventricle even during periods of increased demand, relatively protecting myocardial function in this patient.

This case not only highlights the unusual anatomy of anomalous LCA arising from the mid-RPA but also illustrates an interesting physiological scenario that may have contributed to the relative blunting of cardiac compromise, resulting in a later clinical presentation. This underscores the need for careful evaluation of the coronary artery origins in areas other than the pulmonary root when anomalous connection of a coronary artery is suspected.

Disclosures
None.

References
1. Brooks HSJ. Two cases of abnormal coronary artery of the heart arising from the pulmonary artery; with some remarks upon the effect of this anomaly in producing cirrloid dilation of the vessels. J Anat Physiol. 1885;20:26–29.
Infant With Anomalous Left Coronary Artery Arising From the Right Pulmonary Artery and Ventricular Septal Defect

Ajay K. Bhatia, Joseph Kreeger, Timothy Slesnick, Brian Kogon and Dennis W. Kim

Circulation. 2013;127:e544-e546
doi: 10.1161/CIRCULATIONAHA.112.155226

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2013 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/127/15/e544

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2013/04/12/127.15.e544.DC1