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 postoperative 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 300 000 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 \( \approx 2 \) 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 \( V_4 \) through \( V_6 \) 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 dis-
tress, and fussiness that may obfuscate the diagnosis of con-
gestive 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 sud-
den 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 pul-
monary 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 pul-
monary 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 aris-
ing from the pulmonary artery: with some remarks upon the effect of
this anomaly in producing cirrroid dilation of the vessels. J Anat Physiol.
1885;20:26–29.
2. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left cor-
onary artery from the pulmonary trunk: its clinical spectrum, pathology,
and pathophysiology, based on a review of 140 cases with seven further
3. Neufeld HN, Schneeweiss A. Coronary Artery Disease in Infants and
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

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

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/