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CASE REPORT

BEGINNER

CLINICAL CASE

Echocardiographic Diagnosis of Bland-White-Garland Syndrome in an Asymptomatic Adult

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ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital defect and usually diagnosed within the first 2 months of life. Only 10% of patients survive to adulthood largely in part to the formation of extensive collaterals from the right to left coronary arteries. We present a case of ALCAPA diagnosed in an asymptomatic adult through a transthoracic echocardiogram (TTE). (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1021–4) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENT ILLNESS

A 37-year healthy man presented to establish care with a primary care physician. He had no specific complaints. His vitals were within normal limits with a heart rate of 74 beats/min, blood pressure of 124/60 mm Hg, respiratory rate of 16 breaths/min, and oxygen saturation of 98% on room air. His physical examination was notable for a grade 3 of 6 holosystolic murmur heard best at the apex that radiated to his back.

The patient had a history of measles when he was 2 years of age. Otherwise, his past medical history was unremarkable.

LEARNING OBJECTIVES

- To highlight the importance of understanding the vascular anatomy of ALCAPA, clinical presentation in adults, and diagnosis.
- To identify the echocardiographic features typical of ALCAPA and use in combination with clinical data to significantly reduce misdiagnosis or nondiagnosis, thereby improving survival.
- To understand the post-operative management of patients with ALCAPA.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for an apical holosystolic murmur is mitral regurgitation owing to mitral valve dysfunction.

INVESTIGATIONS

Electrocardiography was notable for a left anterior fascicular block. His laboratory work-up was within

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ABBREVIATIONS AND ACRONYMS

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery

LCA = left coronary artery

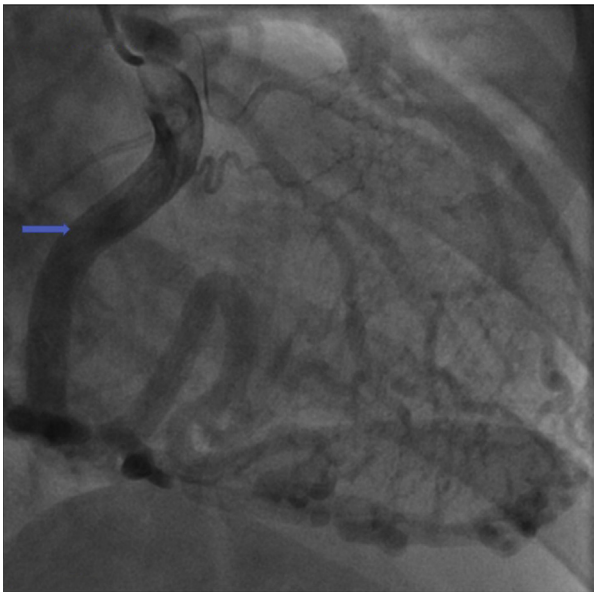
RCA = right coronary artery

normal limits. Transthoracic echocardiography revealed an ejection fraction of 45%, moderate posteriorly directed mitral regurgitation, mild hypokinesis of anteroseptum and anterior, anterolateral, and apical segments of the left ventricle. Color Doppler showed low-velocity, diastolic linear flow in the septum from inferoseptum to anteroseptum, consistent with coronary vessel flow. Low-velocity diastolic flow was also noted into the pulmonary artery, representing reversed flow from an anomalous left main emptying into the pulmonary artery (Videos 1, 2, 3, and 4). Overall findings were highly characteristic of an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). He underwent a stress echocardiogram in which he achieved target heart rate and exercised for 13 min without experiencing chest pain or discomfort. Stress electrocardiography was notable for 3-mm ST-segment depressions in the lateral leads (V_4 to V_6) and 1-mm ST-segment elevation in the aVR lead. Post-exercise echocardiography showed more prominent hypokinesis of the anterior wall and hypokinesis of anteroseptum. Mitral regurgitation remained unchanged from baseline. Cardiac catheterization confirmed the presence of an anomalous left coronary artery (LCA) originating from the pulmonary artery. Right coronary angiography showed a very large right coronary artery (RCA) that filled the entire LCA retrograde via collaterals with flow reversal in the LCA and contrast emptying into the pulmonary artery (**Figures 1 and 2**). Ascending aortography confirmed that there was no LCA arising from the ascending aorta (**Figure 3**). His filling pressures were normal, and there was no step-up in oxygen saturation present.

MANAGEMENT

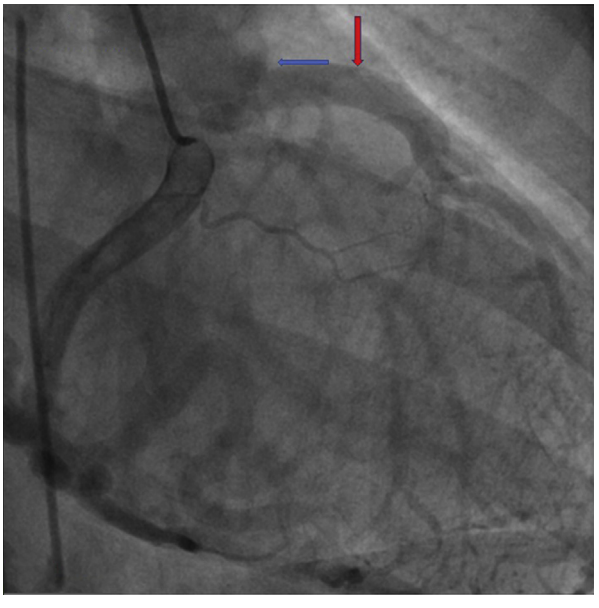
The patient then underwent successful reimplantation of the LCA with concurrent mitral valve repair (**Figures 4 to 6**). His post-operative course remained uneventful, and he was discharged on aspirin and warfarin, given the very robust competing collateral flow with a dilated RCA, which carries a risk of coronary thrombosis.

FIGURE 1 Dilated Tortuous Right Coronary Artery as Seen on Coronary Angiogram



Coronary angiogram **(arrow)** showing the dilated and tortuous right coronary artery with extensive collaterals to the left coronary artery.

FIGURE 2 Filling of Left Coronary Artery Through an Extensive Network of Collaterals From the Right System



Coronary angiogram showing filling of left coronary artery via collaterals **(red arrow)**. The **blue arrow** points to the origin of the left coronary artery from the pulmonary artery.

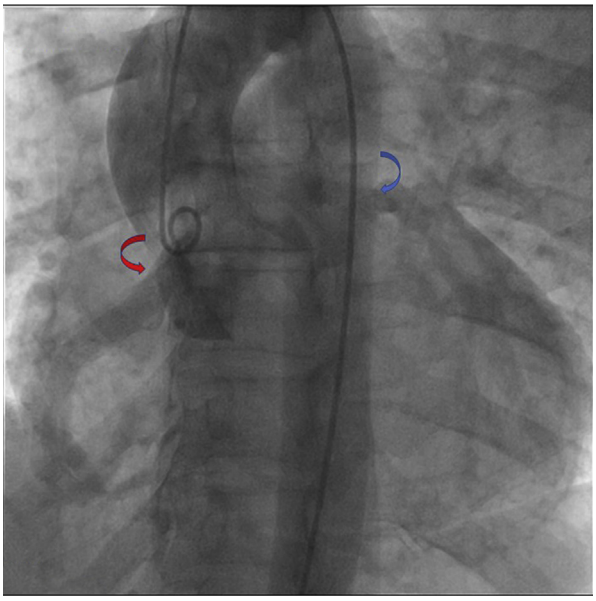
FOLLOW-UP

On his 1-month post-operative follow-up, he reported doing well. He admitted to realizing that he may have had some chest discomfort while climbing hills but that this has completely resolved since his surgery. A murmur was no longer heard on a physical examination. He is compliant with his aspirin and coumadin without bleeding issues.

DISCUSSION

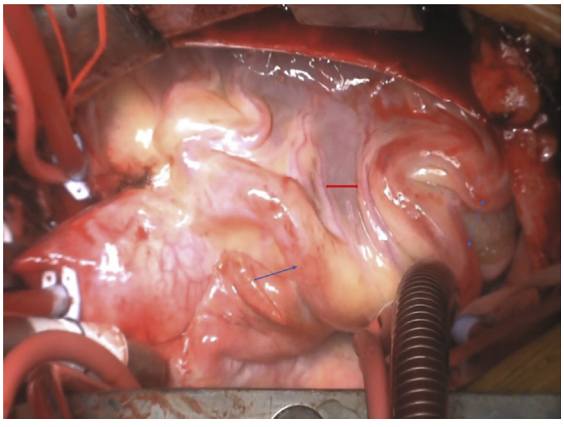
Bland-White-Garland syndrome, also known as ALCAPA, was first described in the literature in 1933 by Bland, White, and Garland (1). ALCAPA is a very rare congenital anomaly that occurs in 1 in 300,000 births, with a 90% mortality within the first year of life when left untreated (2). Although it most commonly presents in infancy, 10% of children with this congenital anomaly will survive to adulthood. Case reports on adults with ALCAPA show that their first presentation is on a spectrum of being asymptomatic to acute heart failure, myocardial ischemia, and sudden cardiac death (3,4). The degree of collateral formation between the RCA and LCA serves as a significant predictor for symptoms. Dilation of the RCA and the extensive collateralization from the RCA to the left system occurs as a compensation for the lower blood oxygen level in the LCA. Continuous blood from the RCA into the LCA and then into the low-pressure pulmonary artery results in coronary steal phenomenon. Papillary muscle and LV lateral wall dysfunction induced by chronic ischemia result in mitral regurgitation (5). Because the incidence of an adult type of ALCAPA is very low, often asymptomatic, and lacking ST-T changes on electrocardiography, patients with this congenital anomaly go through life either undiagnosed or misdiagnosed. Echocardiography is usually the initial diagnostic tool. Echocardiographic features including color Doppler mapping can show a continuous shunt retrograde into the pulmonary artery lumen at the abnormal origin of the LCA. Abundant collateral blood flow may be observed in the myocardium, especially in the interventricular septum. Other secondary features include left ventricular dilation and dysfunction, papillary muscle fibrosis, mitral valve prolapse or regurgitation, abnormal wall motion, and dilatated RCA (5). Although magnetic resonance angiography and computed tomography angiography have been increasingly used for the diagnosis of ALCAPA, these modalities are unable to determine intravascular blood flow conditions and require administration of contrast dye. Echocardiography is an indispensable diagnostic tool because it is noninvasive, is low cost, and has the ability to show the origin of the coronary

FIGURE 3 The Origin of the Left Coronary Artery From the Pulmonary Artery and Right Coronary From the Aorta



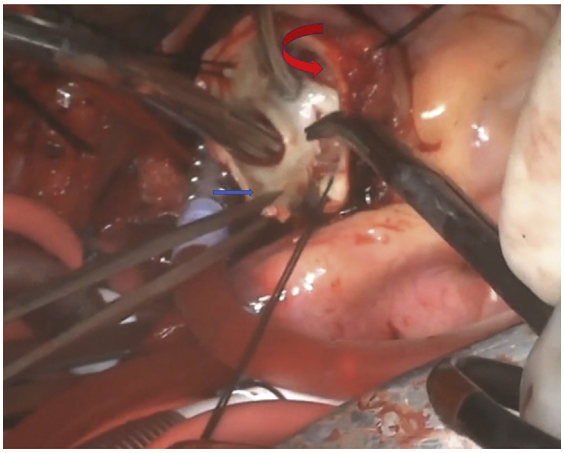
Aortogram shows origin of the left coronary artery from the pulmonary artery **(blue arrow)**. The **red arrow** points to the origin of the right coronary artery from the right coronary sinus aorta.

FIGURE 4 Dilated Tortuous Epicardial Right Coronary Artery as Seen Intraoperatively



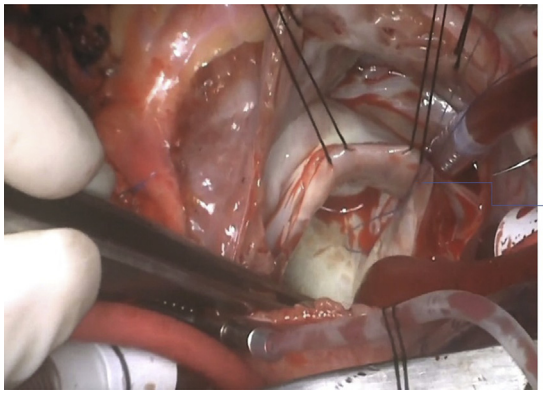
Intraoperative photo showing the tortuous and dilated right coronary artery **(blue arrow)** with extensive collaterals **(red arrow)**.

FIGURE 5 Intraoperative Image of Mitral Valve Repair



Intraoperative photo showing the pulmonary artery **(blue arrow)** being opened and the deep-seated origin of left coronary artery **(red arrow)**.

FIGURE 6 Intraoperative Image Showing Mitral Valve Repair



artery. An obvious drawback is when the echocardiographer does not have comprehensive knowledge of ALCAPA. Of note, the diagnosis may be difficult in the setting of pulmonary hypertension, as the blood flow in the LCA might be forward, and the "stealing blood" sign of the coronary artery is not present.

CONCLUSIONS

ALCAPA is a rare congenital heart disease that carries a high mortality rate and may lead to sudden death if left untreated. An understanding of the typical echocardiographic features of ALCAPA is critical in order to obtain the often elusive diagnosis. Relatively young patients noted to have depressed left ventricular systolic function and concomitant mitral regurgitation without underlying coronary artery disease should raise suspicion for anomalous origin of a coronary artery. Ultrasound imaging with transthoracic echocardiography serves as an essential tool for the early diagnosis of this congenital anomaly. An operator's comprehensive knowledge, diagnostic awareness, and echocardiographic skills are critical for the accurate diagnosis of ALCAPA.

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KEY WORDS ALCAPA, coronary anomaly, dilated coronary artery, echocardiography

APPENDIX For supplemental videos, please see the online version of this paper.