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Multimodality Imaging of Rare Adult Presentation of ALCAPA Treated With Takeuchi Repair

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A 17-year-old African-American female patient was referred to our institution to evaluate an incidentally diagnosed murmur. She lacked stigmata of chronic illness and had no abnormalities on examination other than a flow murmur at the left and right upper sternal borders. She had occasional sharp, left-sided chest pain, occurring at rest and denied dyspnea; however, she had never been active according to her mother, showing little interest in sports or other exertional activities. A transthoracic echocardiogram demonstrated a dilated proximal right coronary artery without clear evidence of a left main ostium. Invasive coronary angiography demonstrated the left coronary artery (LCA) originating from the left posterior sinus of the pulmonary artery (ALCAPA) (**Figures 1A and 1B**, Online Videos 1 and 2) with a very large and ectatic right coronary artery (RCA) with extensive collaterals. Shunt fraction at catheterization was 1.4. For enhanced 3-dimensional orientation of the coronary arteries and great vessels, a computed tomography angiogram was performed that corroborated the catheterization findings (**Figure 1C**). Because of the relatively asymptomatic nature of her presentation, the patient underwent a treadmill exercise test where she was unable to go beyond 10 min and had significant ischemic ST-segment depression (**Figure 1D**). Perfusion imaging showed a large ischemic area in the anterior, anterolateral, and apical myocardium (**Figure 1E**). She subsequently underwent a Takeuchi repair with a 5-mm Gore-Tex baffle (Gore & Associates, Newark, Delaware) because of extensive collateralization and origin of the LCA from the leftward pulmonary sinus (**Figures 1F and 1G**) and has done well since then.

ALCAPA is a rare congenital coronary anomaly that is reported to occur in 1 in 300,000 live births with a mean age at diagnosis of 10.2 months (1). The incidence of death without surgical correction is high, with a reported mortality of 90% in the first year of life (1). Clinically, patients present with congestive heart failure, mitral regurgitation, myocardial ischemia, or sudden death (2). Broadly, patients can be divided into either an infantile presentation or adult presentation. Patients who present as infants often have a small, mildly dominant or nondominant RCA without significant collateral vessels to the left coronary system (2). They present early in life with left ventricular dysfunction due to myocardial ischemia, congestive heart failure, and ischemic mitral regurgitation from papillary muscle dysfunction. Patients who present later into adulthood typically have a large dominant RCA with significant collateral vessels supplying the left coronary system (2). Although adult patients may be asymptomatic for a period of time, they usually present with ischemic symptoms, left ventricular dysfunction, malignant arrhythmias, or sudden death. Surgical re-establishment of the dual coronary system is the preferred management for patients with ALCAPA. There are 2 surgical techniques that are primarily used: LCA translocation and intrapulmonary tunneled repair known as the Takeuchi technique (**Figure 1H**) (3). In cases in

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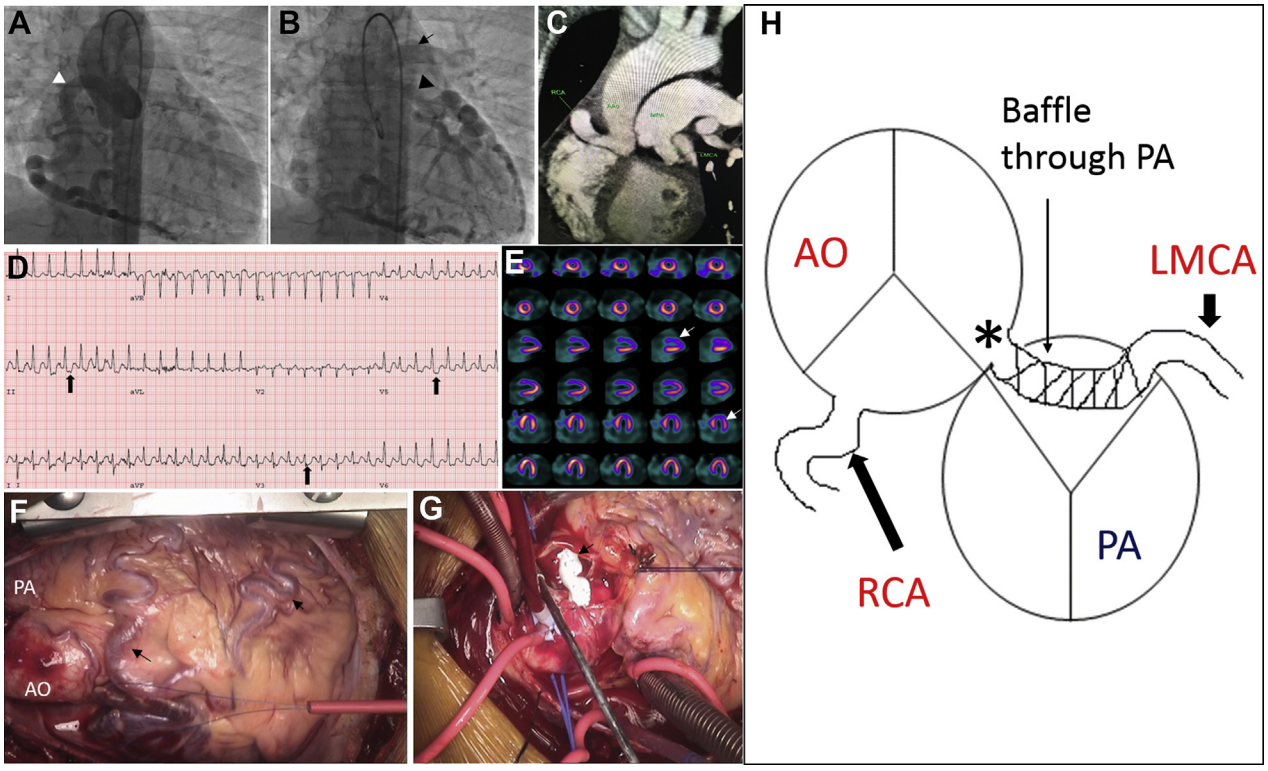
FIGURE 1 **Anomalous Left Coronary Artery From Pulmonary Artery Treated by Takeuchi Repair**



(A and B) Aortogram showing a large right coronary artery **(white arrowhead)** that fills the left coronary artery **(black arrowhead)** via collaterals; the left coronary artery drains into pulmonary artery (PA) **(black arrow)** (Online Videos 1 and 2). **(C)** Computed tomography angiography showing left coronary origin from the posterior sinus of the PA. **(D)** Electrocardiogram after treadmill exercise with marked ST-segment depression **(black arrows)**. **(E)** Stress and rest single-photon emission computed tomography images showing severe, but reversible, perfusion defects in the anterior and lateral walls **(white arrows)**. **(F)** Intraoperative visualization of ectatic right coronary artery (RCA) with dilated collateral vessels. **(G)** Gore-Tex baffle **(black arrow)** placed from the aorta through the PA into the left coronary ostium. **(H)** Schematic cartoon depicting the Takeuchi repair: creation of an aortopulmonary window **(*)** connected to a Gore-Tex baffle traversing the PA anastomosed to the ostium of the left main coronary artery (LMCA). AO = aorta.

which the LCA is short, is relatively fixed due to extensive collateral vessels, or is located too far from the aorta for direct implantation, the Takeuchi technique is favored (2). Indications for reoperation after Takeuchi repair are baffle stenosis, supravalvular pulmonic stenosis, and baffle leak (3).

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KEY WORDS ALCAPA, anomalous coronary artery, Takeuchi repair

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