Anomalous Left Coronary Artery Originating From the Pulmonary Artery Diagnosed in an Adult

Lindsey Croll, MSc, Michael Ellerman, DO, Muhammad J. Ahsan, MD, and Manik Veer, MD,
Des Moines, Iowa

INTRODUCTION

We present a nonatherosclerotic myocardial ischemia and cardiomyopathy in a young woman. An anomalous left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome, is a congenital heart disease that affects 1 in 300,000 births and is very rarely first diagnosed in adulthood.[1,2] Adults present with signs and symptoms of heart failure and ischemia; however, symptoms and presentation depend on the extent of collateralization from the right coronary artery (RCA).[2,3] Our case highlights the physiological challenges presented in adulthood due to this congenital anomaly and their subsequent management.

CASE PRESENTATION

A 40-year-old woman with a medical history of heart failure with reduced left ventricular ejection fraction (LVEF) of 40%, cervical cancer status post chemotherapy and radiation currently in remission, obstructive sleep apnea, obesity, and 1 pack per day tobacco use presented with worsening dyspnea on exertion, intermittent chest pain and fatigue for 2 to 3 weeks, orthopnea, and paroxysmal nocturnal dyspnea. These symptoms initially began 1 year prior with exertion but had recently progressed to symptoms at rest.

Physical examination showed tachypnea with respiratory rate of 22 breaths per minute, bibasilar crackles, jugular venous pressure of 14 cm of water, mild bilateral leg edema, and mild respiratory distress despite 3 L/minute nasal cannula oxygen. Complete blood count, general chemistry, and cardiac enzymes were unremarkable. B-type natriuretic peptide was elevated at 649 pg/mL. Chest x-ray showed bilateral pulmonary congestion and diffuse pulmonary edema. Chest computed tomography angiogram showed no pulmonary embolism and confirmed findings of intrathoracic fluid overload. No overt coronary artery calcifications were noted. Coronary artery origins were not evaluated due to lack of electrocardiogram gating and motion artifact, although an ectatic RCA was visualized. Transthoracic echocardiogram (TTE) demonstrated an LVEF of 40%, mild left ventricular (LV) dilation (LV- internal end-diastolic diameter, of 64 mm; LV internal end-systolic diameter, 52 mm), normal right ventricular size and function, and severe left atrial (LA) dilation (LA volume index, 55 mL/m^2). No regional wall motion abnormalities were documented.

Management was started for acute systolic heart failure. Once the patient was euvolemic, nuclear myocardial perfusion imaging was performed, which showed a large area of anterior, anterolateral, and apical ischemia without evidence of infarction (Figure 1). Coronary angiogram was recommended, but the patient declined due to improvement in symptoms. The patient was lost to follow-up.

Two years later, the patient presented again with dyspnea and chest pain. Coronary angiogram was performed, which demonstrated a large ectatic RCA with collaterals to the left coronary system (Figure 2). During the angiogram, the left main coronary artery (LMCA) was not visualized, and the entire left coronary circulation was visualized via collaterals from the RCA (Video 1). An aortogram was performed that did not reveal the ostium of the LMCA, raising suspicion for anomalous LMCA. On long cine loop, significant contrast was noted in the main pulmonary artery (MPA; Video 1).

A repeat TTE was performed that again demonstrated a mildly dilated left ventricle (LV; LV internal end-diastolic diameter, 58 mm) with an LVEF of 45% (Figure 3), septal wall motion abnormalities (Figure 4), grade II diastolic dysfunction, mild mitral regurgitation, and a severely dilated LA (Video 2). No evidence of patent ductus arteriosus or septal defects was noted.

Coronary computed tomography angiogram (CCTA) demonstrated the origin of the LMCA arising from the MPA, confirming the diagnosis of ALCAPA without significant atherosclerotic coronary artery disease (Video 3, Figures 5-7). For reasons not documented, rather than left coronary reimplantation, they underwent patch ligation of the anomalous LMCA with coronary artery bypass. Grafts included left internal mammary artery (LIMA) to the left anterior descending artery and a vein graft to the obtuse marginal.

DISCUSSION

Anomalous left coronary artery from the pulmonary artery is a rare congenital heart disease that usually ends in failure to thrive and congestive heart failure in infancy, affecting ~1/300,000 newborns in the United States.[1] If left untreated, 90% of patients die during the first year of life due to myocardial ischemia and heart failure.[4] However, adults who survive are found to have significant collateralization from the right to the left coronary artery system. Presenting symptoms typically depend on the degree of collateralization as this determines the severity of ischemia and remodeling.[5,6] Our case highlights several pathophysiological processes that are unique to this congenital anomaly.

From the Iowa Heart Center, MercyOne Des Moines Medical Center, Des Moines, Iowa.

Keywords: ALCAPA, Anomalous, Ischemia, Cardiomyopathy

Correspondence: Michael Ellerman, DO, Iowa Heart Center, MercyOne Des Moines Medical Center, Des Moines, IA, 1111 6th Avenue, Des Moines, Iowa 50314. (E-mail: *michaelellerman09@gmail.com*).

2468-6441
https://doi.org/10.1016/j.case.2023.12.027

VIDEO HIGHLIGHTS

Video 1: Invasive coronary angiography, injection of the RCA, left anterior oblique caudal view, demonstrates the enlarged RCA with collaterals to the left anterior descending artery and then draining into the MPA, diagnostic for ALCAPA.

Video 2: Preoperative two-dimensional TTE, apical 4-chamber view, demonstrates the dilated LV and LA reduced LV systolic function with focal segmental akinesis in the mid inferoseptum.

Video 3: CCTA, thin-slice axial cine-display from superior to inferior obtained preoperatively, demonstrates the left coronary origin from the inferior aspect of the MPA.

View the video content online at www.cvcasejournal.com.

In ALCAPA, a large left-to-right shunt from the RCA to the MPA results in LA and dilation, analogous to a ventricular septal defect. The initial findings of a dilated MPA, dilated and dysfunctional LV, and dilated LA represent the morphological changes consistent with LV volume overload associated with ALCAPA. This represents blood shunting from the RCA, a high-pressure system, into the MPA, a low-pressure system. The right ventricle does not present with morphological changes until there has been significant remodeling secondary to pulmonary hypertension.

Patients who survive childhood usually present in early adulthood secondary to heart failure. This shunt, theoretically, leads to not only LV volume overload but also coronary steal resulting in angina and progressive LV systolic dysfunction.[7]

Our intraoperative transesophageal echocardiogram, done during coronary artery bypass grafting, presents another unique anatomical feature of this syndrome. On the transesophageal echocardiogram, what was suspected to be the left coronary ostium was visualized arising from the left coronary cusp (LCC; Figure 8), although on CCTA, no coronary ostium was visualized from the LCC. This represents a pseudo-ostium caused by acoustic dropout of the LCC and MPA tissue planes giving the impression there is a blind pouch of a rudimentary left main artery from the LCC. On CCTA, the left coronary artery is seen to originate directly from the MPA.

Management is surgical correction, with LMCA reimplantation to the LCC as the standard of care.[8] Surgical ligation of the anomalous LMCA with bypass grafting as in this case has been described in the literature, although this can result in graft longevity complications due to competitive flow from right-to-left collaterals. Given the ectatic RCA with large collaterals to the left coronary system, there is a hypothetical risk of bypass grafts becoming atretic after bypass grafting. Repeat coronary angiography 3 years after our patient's bypass surgery noted obstructive disease in the vein graft but widely patent LIMA graft. We hypothesize that the LIMA graft has higher perfusion pressures than the right-to-left collaterals and hence remains patent. The closure of the venous graft was likely secondary to competitive retrograde flow from the patent LIMA graft. This example further supports left coronary reimplantation rather than bypass grafting of the left system as the standard of care.

The presence of a large ectatic RCA with right-to-left collaterals on coronary angiography along with unexplained LV dysfunction, particularly regional wall motion abnormalities without evidence of atherosclerotic coronary artery disease, should raise the suspicion for ALCAPA. Our patient had a diagnosis of cardiomyopathy for more than a decade which, due to their young age, was thought to be nonischemic in etiology and was attributed to chemotherapy for cervical cancer. Thus, differential diagnosis of various forms of anomalous coronary arteries should be considered in patients with early-onset heart failure, LV systolic dysfunction and dilation, and myocardial ischemia. The average age of presentation in adults is 41 years old and is complicated by pulmonary hypertension, ventricular arrhythmias, heart failure, and sudden cardiac death if not diagnosed and treated.[4]

CONCLUSION

Anomalous left coronary artery from the pulmonary artery presenting in adulthood is a rare diagnosis. The diagnosis should be on the differential diagnosis of any young patient presenting with left-sided chamber dilation, LV dysfunction, and angina. Diagnosis is confirmed through multimodality imaging including angiography, TTE, CCTA and cardiovascular magnetic resonance imaging. Management is surgical reimplantation of the LMCA to the aorta in addition to guideline therapy for heart failure.[4]

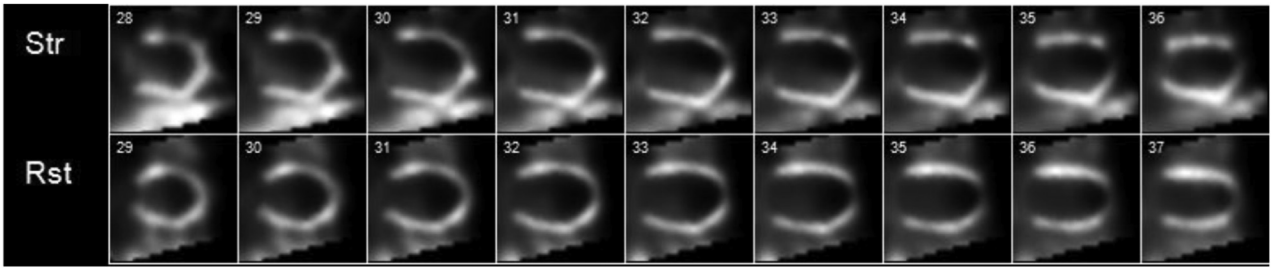

Figure 1 Vasodilator (regadenoson) stress (*top row*) and rest (*bottom row*) technetium myocardial perfusion imaging single-photon emission computed tomography imaging, vertical long-axis orientation, demonstrates a dilated LV with a large reversible perfusion deficit in the mid-distal anteroseptal, anterior, anterolateral, and apical walls.

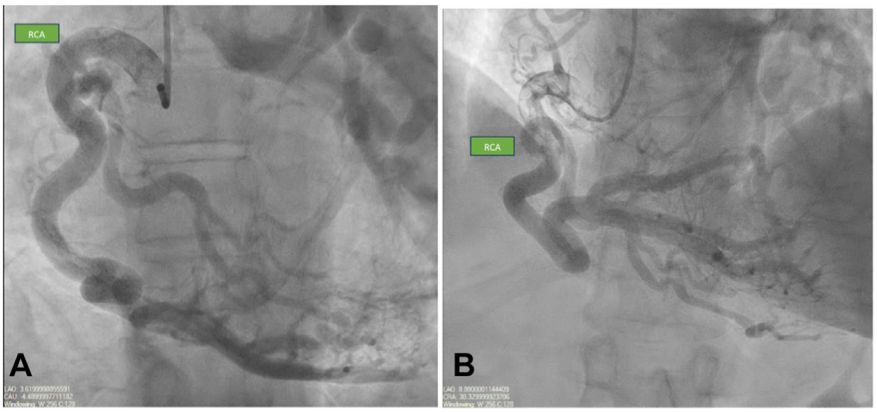

Figure 2 Invasive coronary angiography, injection of the RCA, left anterior oblique caudal **(A)** and cranial **(B)** views, demonstrates the enlarged RCA with collaterals to the left anterior descending artery.

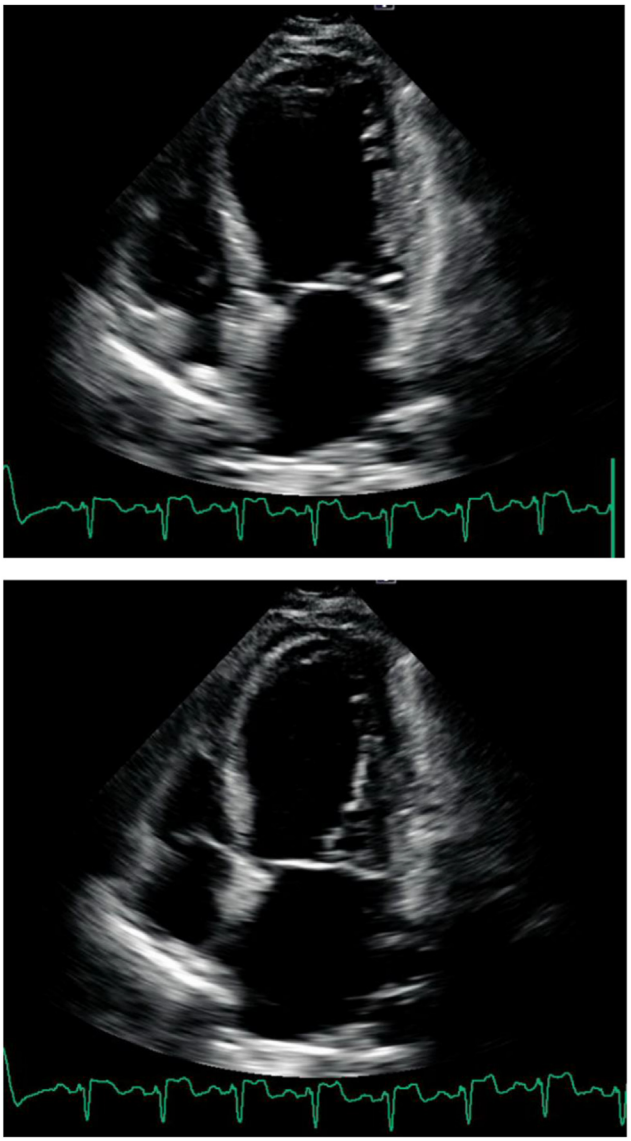

Figure 3 Preoperative two-dimensional TTE, apical 4-chamber view, in end diastole (*top*) and end systole (*bottom*), demonstrates the dilated LV and left atrium with reduced LV systolic function.

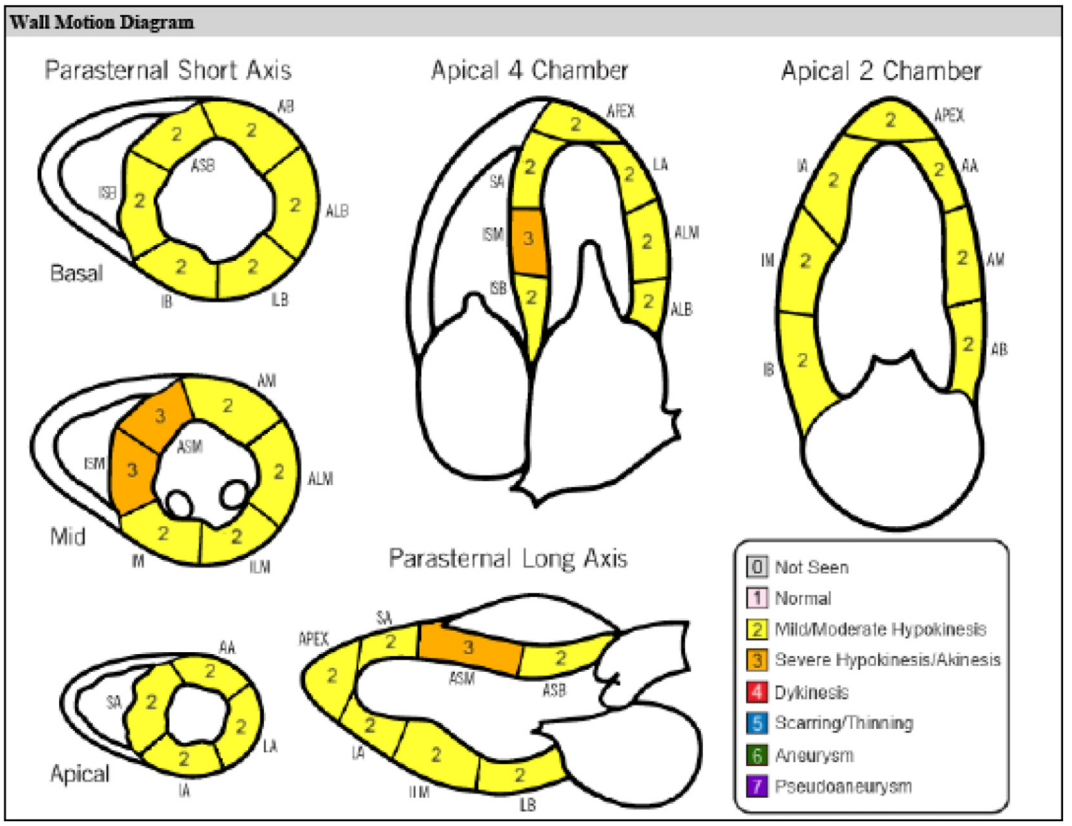

Figure 4 Regional wall motion abnormalities reported prior to the diagnosis of ALCAPA emphasizing the predominant global dysfunction with small focal akinetic regional wall segments.

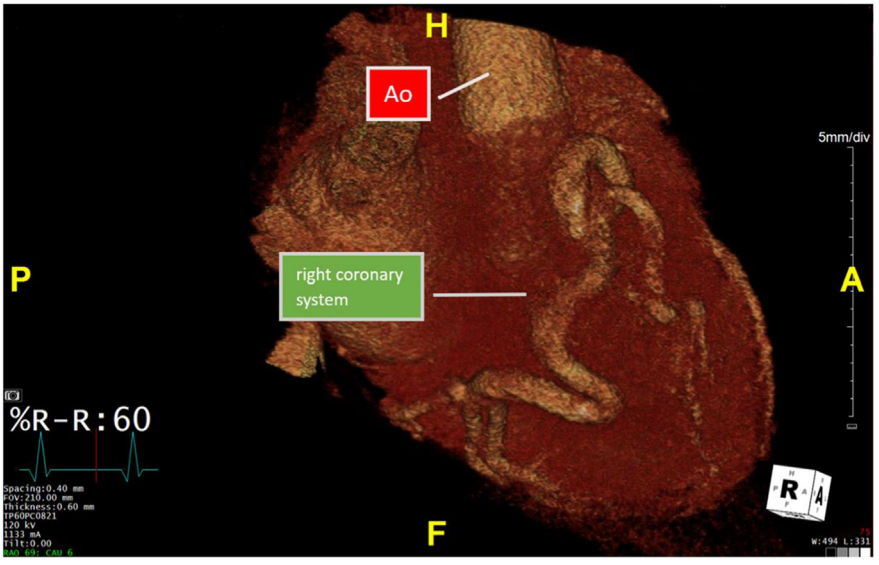

Figure 5 Three-dimensional CCTA, whole-heart volume-rendered reconstruction obtained preoperatively, demonstrates the aneurysmal RCA. *Ao*, Aorta.

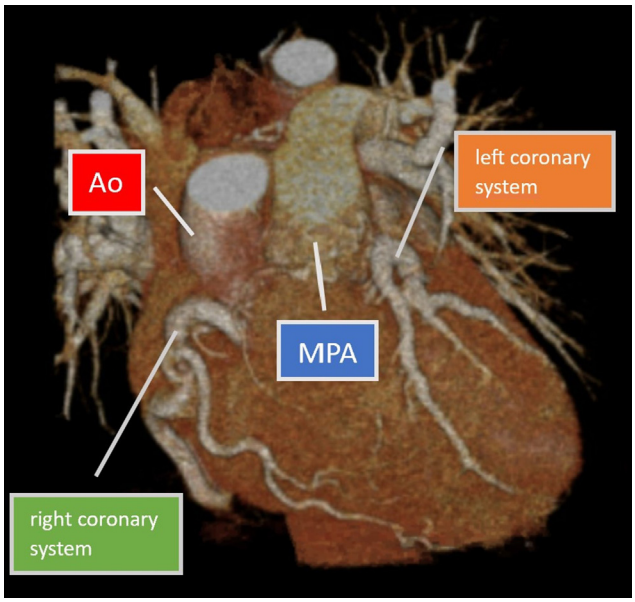

Figure 6 Three-dimensional CCTA, whole-heart volume-rendered reconstruction obtained preoperatively, demonstrates the anomalous left coronary artery originating from the MPA. *Ao*, Aorta.

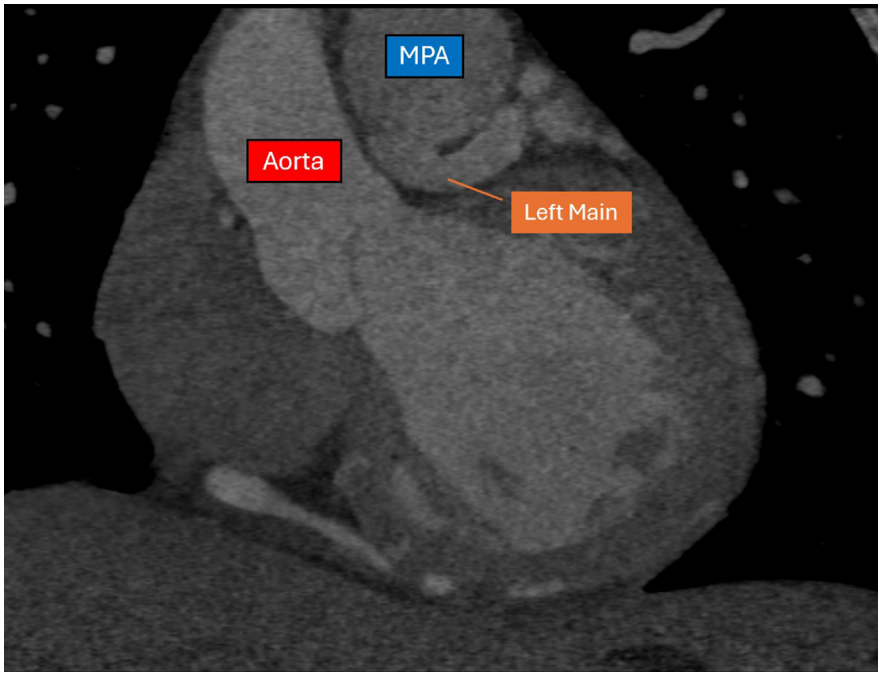

Figure 7 CCTA, multiplanar reconstruction display obtained preoperatively, demonstrates the left coronary origin from the inferior aspect of the MPA.

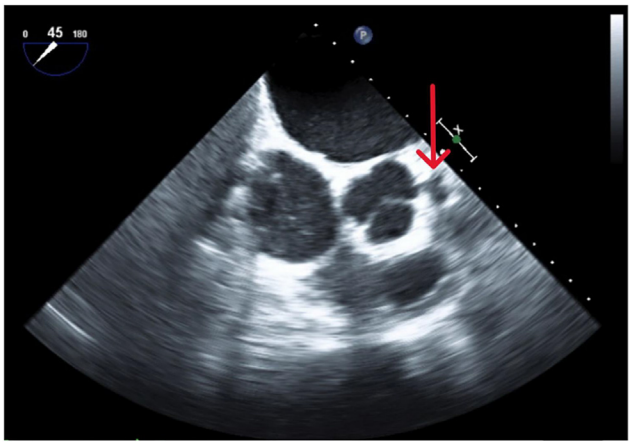

Figure 8 Two-dimensional transesophageal echocardiogram, aortic valve short-axis (45°) view in diastole, demonstrates acoustic dropout between the origin of the LMCA and the aortic root and resulting in the risk of misinterpreting that the ostium of the LMCA arises from the aorta.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.027.

REFERENCES

1. Yau JM, Singh R, Halpern EJ, Fischman D. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. Clin Cardiol 2011;34:204-10.
2. Barbetakis N, Efstathiou A, Efstathiou N, Papagiannopoulou P, Soulountsi V, Fessatidis I. A long-term survivor of Bland-White-Garland syndrome with systemic collateral supply: a case report and review of the literature. BMC Surg 2005;5:23.
3. Cabrera AG, Chen DW, Pignatelli RH, Khan MS, Jeewa A, Mery CM, et al. Outcomes of anomalous left coronary artery from pulmonary artery repair: beyond normal function. Ann Thorac Surg 2015;99:1342-7.
4. Quah JX, Hofmeyr L, Haqqani H, Clarke A, Rahman A, Pohlner P, et al. The management of the older adult patient with anomalous left coronary artery from the pulmonary artery syndrome: a presentation of two cases and review of the literature. Congenit Heart Dis 2014;9:E185-94.
5. Trevizan LLB, Nussbacher A, Da Silva MCB, Ishikawa WY, de Oliveira SA, Sitta MDC, et al. Anomalous origin of the left coronary artery pulmonary artery as a rare cause of left ventricular dysfunction. Circ Cardiovasc Imaging 2019;12:e009724.
6. Zacharias M, Chandok D, Tighe D. A late presentation of an anomalous left coronary artery originating from the pulmonary artery (ALCAPA): a case study and review of the literature. J Cardiol Cases 2015;11:56-9.
7. Napoli F, Aleman R, Brozzi NA, Montero O, Balzan J, Sheffield C, et al. Alternative surgical repair: anomalous left coronary from the pulmonary artery syndrome in adulthood. Ann Thorac Surg Short Rep 2023;1:404-6.
8. Lange R, Vogt M, Hörer J, Cleuziou J, Menzel A, Holper K, et al. Long-Term results of repair of anomalous origin of the left coronary artery from the pulmonary artery. Ann Thorac Surg 2007;83:1463-71.