Life long surveillance is warranted as coronary artery remodels variably after treatment of adult patients with anomalous left coronary artery origin from pulmonary artery

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ABSTRACT

Anomalous left coronary artery origin from pulmonary artery causes heart failure in infancy from ischemia and secondary mitral regurgitation. Rich intramyocardial collateralization may permit survival to adult age, where coronaries become tortuous and aneurysmally dilated. Surgery in adults involves left coronary ligation and providing a bypass graft to the left system, unlike coronary translocation adopted in infants. Unfavorable coronary remodeling in operated adults may lead to late coronary thrombotic occlusions. Two adults with markedly dilated tortuous coronary arteries showed variable remodeling after corrective intervention that impacted outcomes on follow-up. We stress the need for lifelong angiographic surveillance in older patients.

Keywords: Adult congenital heart disease, aneurysmal coronary dilation, computed tomography, coronary angiography, coronary anomaly, coronary thrombosis

INTRODUCTION

Anomalous left coronary artery origin from pulmonary artery (ALCAPA) often presents with heart failure in infancy, but rare adult survivors are prone to sudden death. While 90% of patients succumb in infancy, adult survival is related to rich intramyocardial collateralization from the right coronary artery to the left system.[1] Recent availability of advanced noninvasive cardiac imaging has resulted in increase in their detection. Myocardial ischemia either from coronary steal into the pulmonary artery or thrombotic occlusion of tortuous aneurysmally dilated coronaries may lead to unfavorable outcomes in adults. Dual coronary perfusion is surgically established in adults by left coronary ligation and bypass graft to the left system through arterial or venous conduits.[2,3] Size discrepancy between conduits and dilated coronaries challenges identification of the correct conduit. In one-fourth of patients, left coronary ligation alone is performed without bypass graft to provide a single coronary perfusion.[4] While reverse remodeling of minimally dilated coronaries in infancy is common, a similar reduction in tortuous aneurysmal vessels in adults may not occur. Coronary imaging in two adult patients demonstrates the variations in their unnatural history after their corrective intervention.

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CASE REPORTS

Patient 1

A 38-year-old female with exertional dyspnea was diagnosed to have ALCAPA and preserved left ventricular function. Computed tomography showed aneurysmal

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tortuous proximal right coronary and left anterior descending coronary arteries that measured more than 8 mm. As myocardial perfusion scan demonstrated a reversible ischemia, transcatheter device occlusion of left main coronary artery was performed using a 8 mm muscular ventricular septal occluder to stop the coronary steal as an alternative to surgery after informed consent.[5] Despite having a single coronary perfusion, she remained asymptomatic with improved ejection fraction and regional longitudinal strain on speckle tracking. Exercise testing did not show ischemia on technetium sestaMIBI myocardial perfusion scan after 1 year. Computed tomography after 30 months showed significant reduction in size and tortuosity of the coronary arteries, indicating favorable remodeling [Figure 1]. Multiplanar reconstructed images showed reduction in size in all the coronary artery segments [Figure 2]. Dual antiplatelet medications were downgraded to aspirin after 30 months.

Patient 2

Another 47-year-old female with ALCAPA, aneurysmal coronary dilatation, and preserved left ventricular function underwent surgical ligation of left main coronary ostium and saphenous venous graft to left anterior descending coronary artery 8 years ago establishing dual coronary perfusion. She was treated with aspirin and rivaroxaban as tomographic images showed aneurysmally enlarged coronary arteries. As a second computed tomography after 6 years of surgery showed persistent aneurysmal right coronary artery dilatation more than 8 mm, both drugs were not discontinued [Figure 3]. There were no luminal irregularity, thrombus, or calcific plaques in this tomographic study. She was recently diagnosed to have diabetes mellitus. When she presented 8 years after surgery with unstable angina at rest with elevated troponin levels and nondescript electrocardiographic changes, coronary angiogram showed a total thrombotic occlusion of proximal right coronary artery and proximal anastomotic stenosis of the saphenous venous graft to the left system. Angiogram from the stenotic venous graft collateralized right coronary territory, but inadequate to relieve rest angina. There were multiple focal irregularities in different coronary artery segments [Figure 4]. She underwent a second bypass surgery with three new venous grafts to left anterior descending, posterior descending, and posterolateral

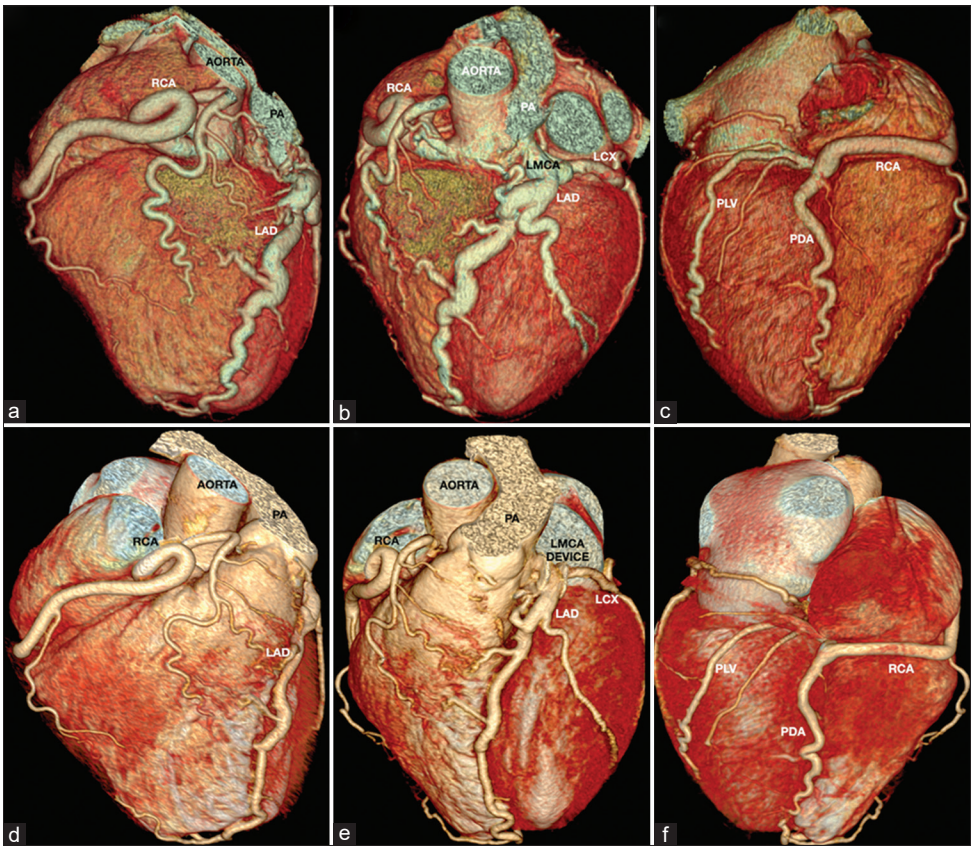
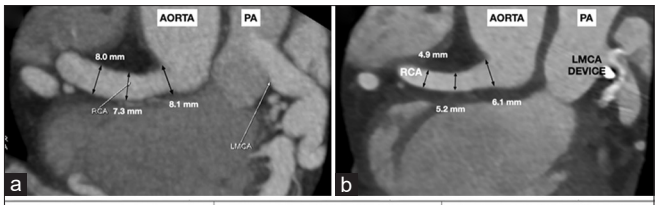


Figure 1: Volume rendered computed tomographic angiography images (a-c) shows anomalous LMCA origin from PA and marked aneurysmal dilation of RCA and LAD artery. The LCX, PDA and PLV branch were also dilated. After 30 months of device occlusion of the left main coronary artery, corresponding images (d-f) show significant reduction in size and tortuosity of all arterial branches. LMCA: Left main coronary artery, PA; Pulmonary artery, RCA: Right coronary artery. LAD: Left anterior descending, LCX: Left circumflex, PDA: Posterior descending artery, PLV: Posterolateral ventricular



Vessel	Pre procedural diameter (mm)	Post procedural diameter (mm) (30 months)
Proximal LAD	10.8	6.5
Mid LAD	8.4	4.7
Distal LAD	4.9	2.4
Proximal LCX	5.1	2.9
Mid LCX	5.2	2.9
Ostia RCA	8.1	6.1
Proximal RCA	8	4.9
Mid RCA	6.9	3.7
Distal RCA	5.8	3.6

Figure 2: Individual coronary dimensions were measured in the preprocedural computed tomography (a) and at 30-month follow-up tomography (b) to study the reduction in the size of corresponding segments in the two studies

ventricular coronary branches. She was continued on aspirin and apixaban.

DISCUSSION

Unnatural history of congenital heart diseases is vital to predict problems that may arise after intervention and plan the appropriate imaging surveillance. Even though the coronary arteries are aneurysmally dilated and tortuous in unoperated adults with ALCAPA, the large blood flow from right coronary artery to the left system prevents thrombosis in a majority permitting their anecdotal survival even up to eighth decade. After arrest of the coronary steal by left coronary ligation, sluggish blood flow in the aneurysmally dilated vessels has been attributed for adverse clinical outcomes proven on autopsy in the operated patients.[6] The slow flow often warrants a precise anticoagulant or antiplatelet strategy. Progressive reduction of the coronary artery size will be a favorable remodeling, but persistent dilatation with thrombotic occlusion will lead to adverse events in the unnatural history of these operated adults. The preinterventional coronary artery dimensions were around 8–10 mm and similar in both adult patients described in this report.

Favorable remodeling involves reduction in the dilatation and tortuosity of the coronary vessels.[7] In the first patient, transcatheter occlusion of left coronary ostium provided a functionally similar result to left main coronary surgical ligation that is adopted in one-fourth of adult surgical patients around the world.[2-4] Serial imaging showed a progressive reduction in the size of the coronary arteries and reduction in the luminal irregularity in this patient that permitted downgradation of antiplatelet medications after 3 years. A similar transcatheter strategy of occlusion of the left coronary

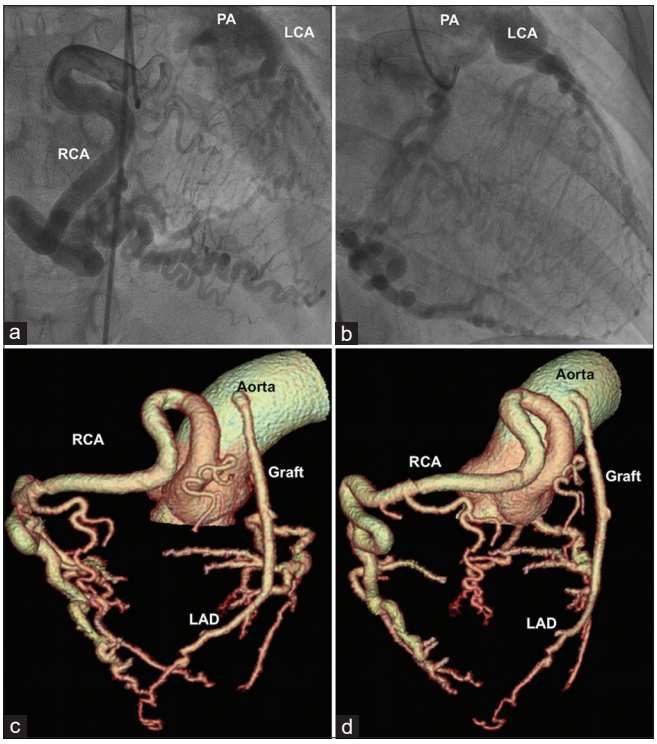


Figure 3: Coronary angiography (a and b) show aneurysmally dilated RCA and late filling of PA trunk through the LCA. Six years later, surveillance computed tomography (c and d) showed persistent aneurysmal RCA dilatation and patent venous graft to LAD artery, necessitating continuation of aspirin and rivaroxaban. RCA: Right coronary artery, PA: Pulmonary artery, LCA: left coronary artery, LAD: Left anterior descending

artery was previously reported.[8] In contrast to the first patient, computed tomography after 6 years in the second patient showed persistent irregular dilation of coronary arteries that did not allow withdrawal of aspirin and rivaroxaban. Despite regular anticoagulation, right coronary artery thrombosis resulted in rest angina and warranted a second bypass surgery.

While 60%–80% of operated adults get dual coronary perfusion either by coronary bypass grafts or translocation, graft occlusions are more common in these patients due to size mismatch between the conduit and dilated coronaries.[2,3,5] Alternative conduits such as turned down subclavian artery, vascular graft from carotid artery has been explored in these dilated coronaries. Creation of watershed zones between the two coronary systems after surgical re-establishment of dual coronary perfusion in adult ALCAPA patients may induce thrombosis in the slow-flow dilated coronary segments. Occlusions of coronary arteries or grafts may remain silent till they lead to sudden death during stress or may present with unstable angina. Serial angiographic surveillance is mandatory to identify silent coronary occlusions as well as progressive thrombotic narrowing. This will permit early management decisions in the operated adults.

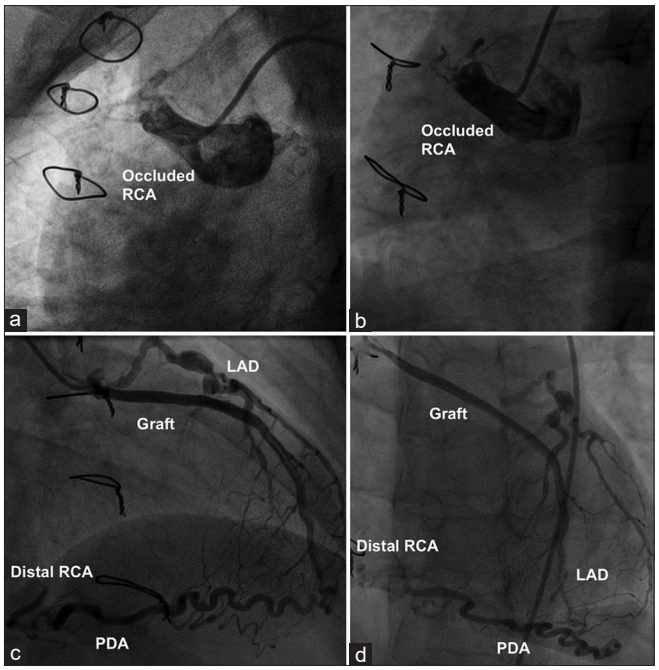


Figure 4: Coronary angiographic evaluation of rest angina showed complete thrombotic occlusion (a and b) of proximal RCA. The narrowed ostium of the saphenous venous graft to LAD artery led to insufficient collateralisation (c and d) of the PDA and distal RCA. RCA: Right coronary artery, LAD: Left anterior descending, PDA: Posterior descending artery

CONCLUSIONS

Variable arterial remodeling in adults warrants their lifelong surveillance, as aneurysmally dilated tortuous coronary arteries may be prone for sluggish flows and thrombosis. Despite establishing dual coronary myocardial perfusion, thrombotic occlusion of vessels or stenosis of grafts may lead to late ischemia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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