Cardiac Computed Tomography in the Diagnosis of Adult Anomalous Left Coronary Artery from the Main Pulmonary Artery in the Caribbean

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ABSTRACT

Definitive diagnosis of an anomalous coronary artery from the main pulmonary artery (ALCAPA) can be difficult. Cardiac computed tomography (cardiac CT) has the ability to safely, cheaply and non-invasively allow direct visualization of the anomalous coronary connection and can demonstrate the systemic-to-venous flow which underlies the coronary steal phenomenon. Here we present the first case of ALCAPA syndrome diagnosed with cardiac CT in Trinidad and Tobago.

Keywords: ALCAPA syndrome, adult congenital heart disease, cardiac CT, Caribbean, Trinidad and Tobago

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INTRODUCTION

Origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly occurring in 1 in 300 000 births. Without surgical correction only 10% of patients survive to adulthood (1). Intermittent myocardial ischaemia during periods of increased activity may lead to left ventricular dysfunction, secondary mitral valve incompetence, malignant ventricular arrhythmias and sudden cardiac death.

CASE REPORT

A 21-year-old male presented to the accident and emergency department with a complaint of recent onset atypical left-sided chest pain of moderate intensity and a history of recurrent syncope since the age of 13 years. Syncope typically occurred with moderate exertion such as running or bicycling and was reported to last five to ten minutes.

Physical examination and vital signs were unremarkable and all laboratory tests were within normal, including troponin assays. Twelve-lead electocardiography showed ST-segment elevation (which met criteria for 'early repolarization' pattern) and left atrial enlargement while chest X-ray suggested cardiomegaly. A provisional diagnosis of hypertrophic obstructive cardiomyopathy was made and the patient was admitted for cardiac monitoring and urgent echocardiography.

Trans-thoracic echocardiography revealed mild left ventricular (LV) dilation with a reduced ejection fraction of 45–50% and moderately elevated estimated LV filling pressure. Moderate tricuspid valve regurgitation and moderate-to-severe mitral valve regurgitation with very severe left atrial enlargement were documented. An estimated right ventricular systolic pressure of 71 mmHg suggested severe pulmonary hypertension. Trans-oesophageal echocardiography confirmed transthoracic findings and also revealed bi-leaflet mitral valve redundancy and prolapse with moderate-to-severe incompetence (estimated effective regurgitant orifice area 0.27 square centimetres and regurgitant volume 34 millilitres). Colour flow doppler and bubble contrast injection did not reveal evidence of intra-cardiac shunt.

Coronary angiography was attempted but the left main (LM) coronary artery could not be engaged. Contrast injection of the right system revealed extensive right-to-left collaterals filing the left anterior descending and left circumflex coronary arteries retrogradely. A conclusion of likely congenitally anomalous take-off of the

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LM coronary with possible complete occlusion of the LM was made. The patient was referred for cardiac computed tomography in order to identify the origin of the LM coronary artery and assess for its possible occlusion.

Cardiac computed tomographic angiography (CTA) demonstrated cardiomegaly and coronary dilation (Fig. 1) and revealed an anomalous origin of the LM coronary artery from the main pulmonary artery [MPA] (Figs. 2–4, 6).

The left anterior descending (LAD) and circumflex (Cx) coronary arteries lay in anatomically typical positions but were both dilated. The right coronary artery (RCA) arose from the non-coronary sinus of the aortic root adjacent to a separate conus ostium and was dilated (Fig. 5).

It supplied collateral branches to the body of the LAD and terminal LAD (Fig. 7) and to the Cx *via* left

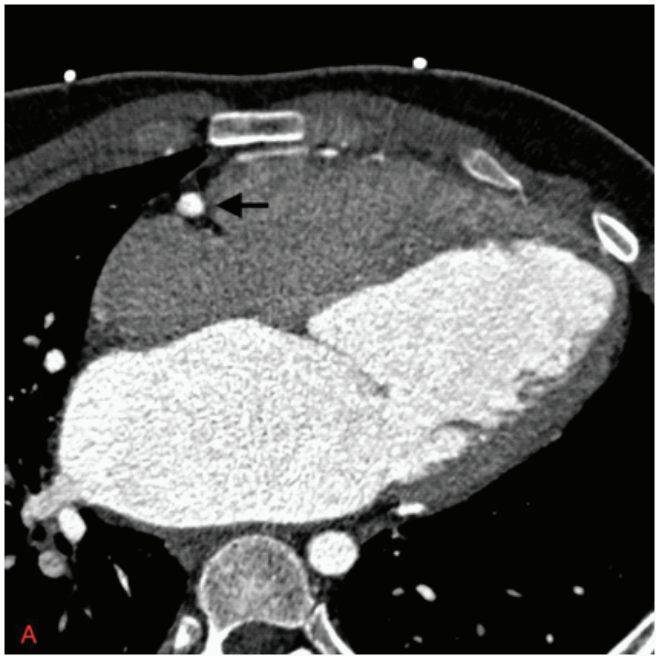

Fig. 1: Cardiomegaly and right coronary artery dilation (arrow).

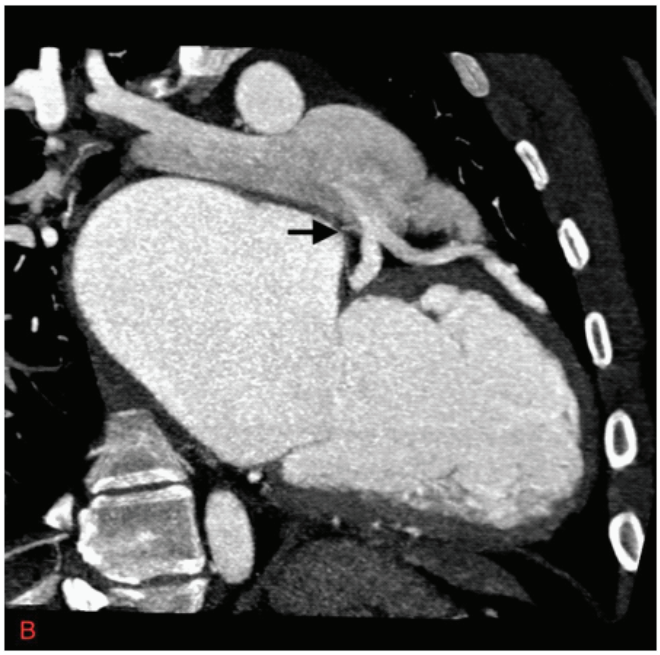

Fig. 2: Left main dividing into left anterior descending and circumflex coronary arteries (arrow).

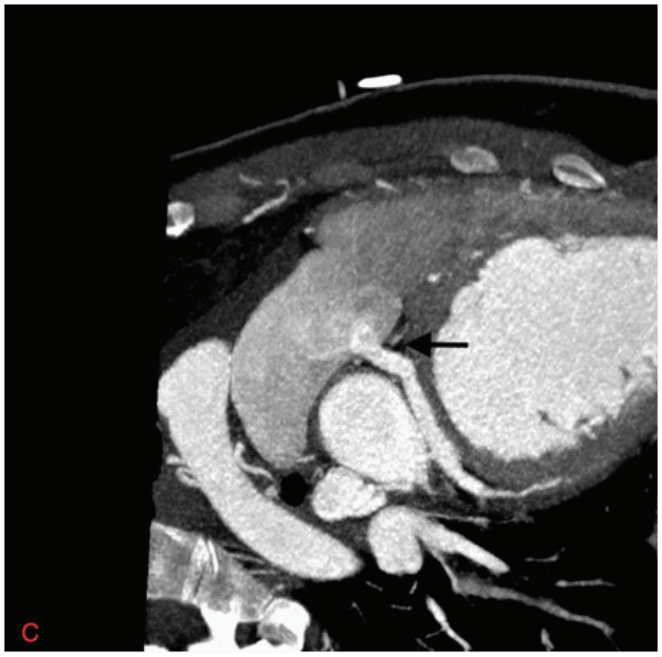

Fig. 3: Contrast flow from left main into main pulmonary artery.

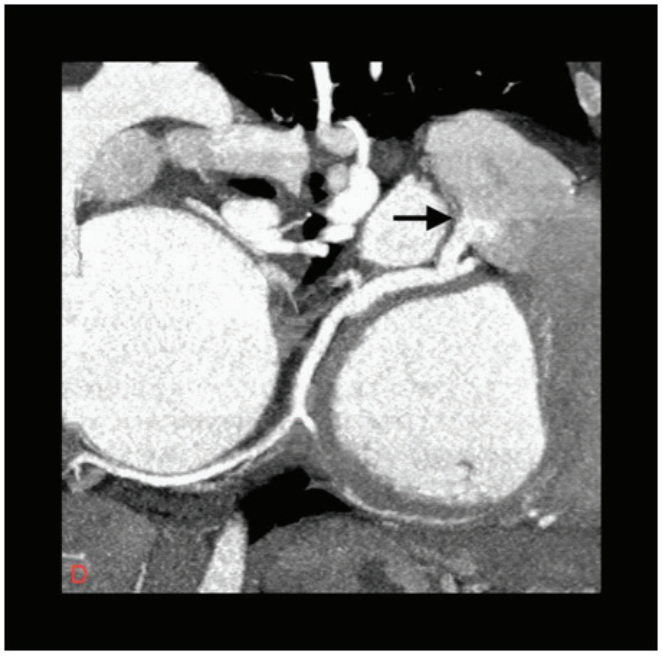

Fig. 4: Contrast flow into main pulmonary artery above the pulmonary valve.

postero-lateral (PL) branches (Fig. 8). Flow of contrast from the LM to the MPA was nicely demonstrated (Figs. 2–4).

DISCUSSION

Dilation of all three coronary arteries resulted from 'left-to-right' shunting of systemic oxygenated blood to the deoxygenated blood of the right-heart circulation. This unique form of arteriovenous fistula resulted in a coronary steal phenomena with myocardial ischaemia, left ventricular dysfunction and dilation and likely malignant ventricular arrhythmias. It seems plausible that this patient's syncopal episodes during moderate exertion represented aborted sudden cardiac death. The additional findings of moderate-to-severe mitral valve regurgitation and severe pulmonary hypertension suggest that prompt surgical repair is prudent.

Fig. 5: Lone right coronary artery from non-coronary sinus.

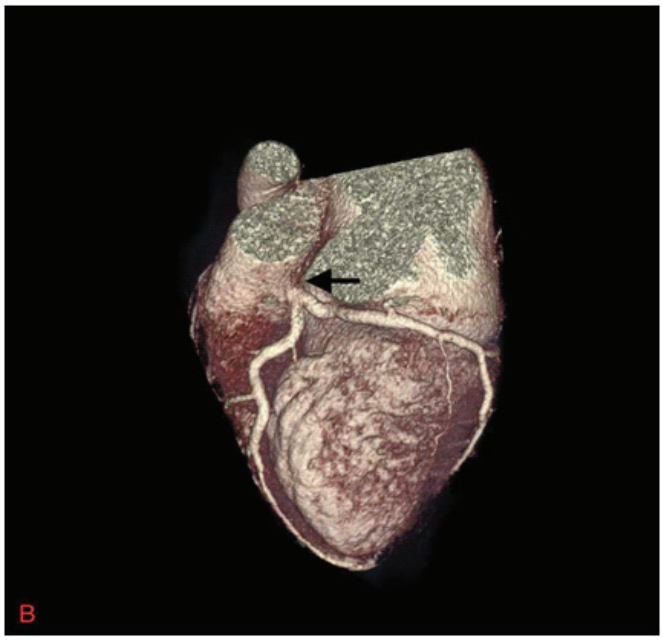

Fig. 6: Left main originating from the main pulmonary artery an dividing into left anterior descending and circumflex coronaries.

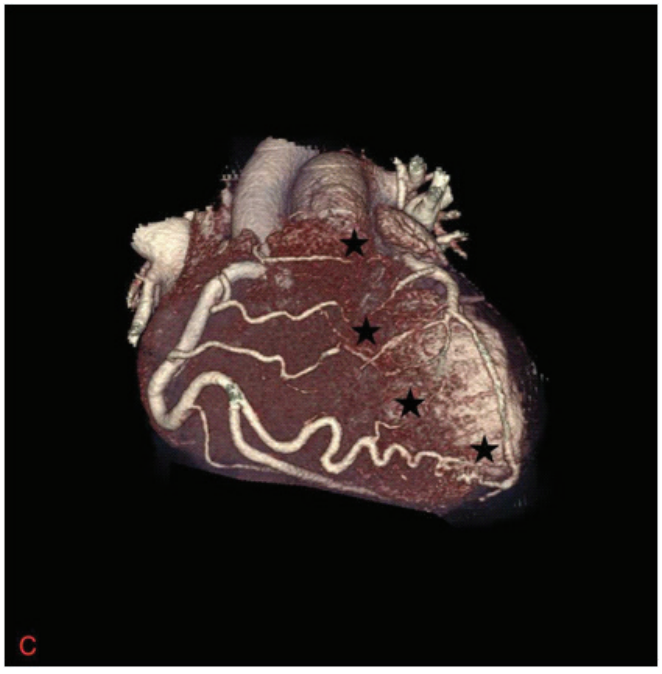

Fig. 7: Collateral circulation from right coronary artery to left anterior descending coronary.

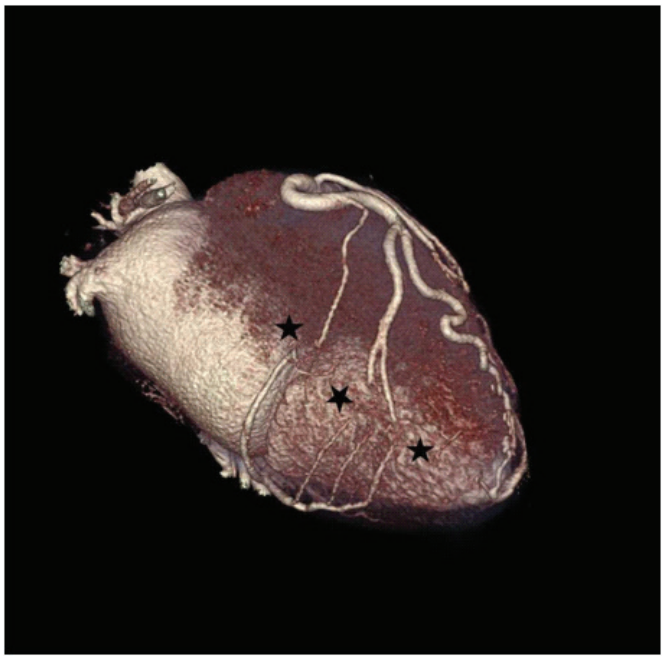

Fig. 8: Collateral circulation from right coronary artery to circumflex coronary.

Restoration of a two-coronary-artery system using either a left subclavian artery or saphenous vein bypass graft to the LM coronary artery with ligation of the anomalous LM connection to the MPA is the preferred approach in adults (2). Mitral valve reconstruction at the time of surgery would also have to be considered in this case as the degree of myocardial improvement and subsequent decrease in valvular incompetence are difficult to predict.

This is the first report in Trinidad and Tobago of a definitive diagnosis of ALCAPA syndrome using cardiac computed tomographic angiography (CTA). There was direct visualization of the LM coronary artery arising anomalously from the main pulmonary artery (MPA) and demonstration of contrast flow from the LM to the MPA. Cardiac CTA is a safe, non-invasive and relatively inexpensive means of assessing suspected complex coronary anomalies. It has the advantage of providing detailed three-dimensional images for cardio-thoracic surgical planning.

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AUTHORS' NOTE:

Dr Bruce Bird designed the cardiac CT protocol for this patient, supervised heart rate reduction, reconstruction of images and reporting of the cardiac CT. He drafted this manuscript as primary author.

Dr Shari Khan was the patient's admitting cardiologist, reported the trans-thoracic echocardiogram and performed and reported the trans-oesophageal echocardiogram. She edited the article as second author.

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