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A case of a 42-year-old patient with anomalous origin of the left main coronary artery from the pulmonary artery who delivered three times with no complications: presentation, diagnosis, and review

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Summary

An anomalous origin of the left coronary artery (LCA) from the pulmonary artery or Bland-White-Garland (BWG) syndrome is a rare congenital cardiac anomaly, which is unusual to survive to adulthood if left uncorrected. The authors report an unusual case of a multiparity patient who delivered several times without any complications presenting at the age of 42 years with recurrent dyspnea on exertion in which echocardiographic findings of diastolic flow near the origin of the pulmonary valve were suggestive of anomalous origin of the LCA. A CT angiography confirmed that the left main coronary artery arises from the undersurface of the pulmonary flow close to its origin and gives rise to a left anterior descending (LAD) and left circumflex arteries.

Key words: Congenital heart disease; Uncomplicated pregnancy; Coronary anomalies; Echocardiography; Computed tomography angiography.

Introduction

An anomalous origin of the left coronary artery (LCA) from the pulmonary artery or Bland-White-Garland (BWG) syndrome is a rare congenital cardiac anomaly, which is unusual to survive to adulthood if left uncorrected. Collaterals to the left system are the key point for the survival to adulthood. CT angiography remains the best modality for diagnosis.

The authors report an unusual case of a multiparity patient who delivered several times without any complications presenting at the age of 42 years with recurrent dyspnea on exertion in which echocardiographic findings of diastolic flow near the origin of the pulmonary valve were suggestive of anomalous origin of the LCA.

Case Report

History dates back to the year 2003, where a 31-years-old female that was previously healthy, except for a history of mitral valve prolapse (MVP) and regurgitation corrected with a mitral ring repair in year 2000, presented for a routine follow up where echocardiography showed a suspicion for a coronary ventricular fistula. Patient was admitted for cardiac catheterization and CT angiography that showed patent right coronary artery (RCA) with prominent vessels in the anterior mediastinum that may be related to the RCA; images were suggestive of a stenosis at the origin of the Left coronary artery (LCA).This was followed by a magnetic resonance angiography (MRA) of the LCA which showed at that time, normal origin of the LCA from the aortic cusps with severe stenosis at its origin. Due to the fact that the patient had normal exercise tolerance and no angina symptoms, no surgical intervention was done and patient was stabilized on medical therapy.

Preceding this symptomatic period patient became pregnant twice with an additional pregnancy after that at the age of 34; all delivered with normal vaginal delivery, with no complications during gestation, labor or postpartum.

Patient remained well without functional limitation and no symptoms until 2014, at the age of 42 where she complained of palpitations and progressive dyspnea in a periphery hospital, EKG showed rapid rate atrial fibrillation that was electrically cardioverted to sinus rhythm.

Patient then presented to the present hospital complaining of recurrent dyspnea on exertion with no symptoms of angina. Echocardiography was done and it showed moderate dilatation of the left ventricle (LV) with mild global LV hypokinesia and an ejection fraction of 45-49%. During diastole, evident Doppler signals were seen near the origin of the pulmonary valve suggestive of aberrant origin of the left main coronary artery from the pulmonary artery (PA) (Figure 1). A CT angiography of the coronary arteries was performed and demonstrated that the left main coronary artery arose from the undersurface of the pulmonary flow close to its origin and gave rise to an LAD and left circumflex arteries (Figures 2, 3). Also it showed the anastomosis between the right posterior descending CA and the LAD (Figure 4).

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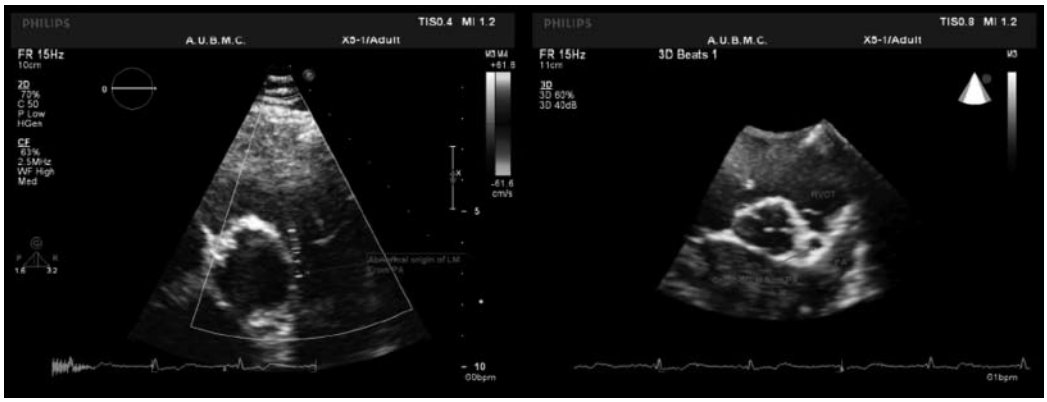

Figure 1. — Echocardiography showing moderate dilatation of the left ventricle (LV) with mild global LV hypokinesia with evident Doppler signals near the origin of the-pulmonary valve suggestive of aberrant origin of the left main coronary artery from the pulmonary artery (PA).

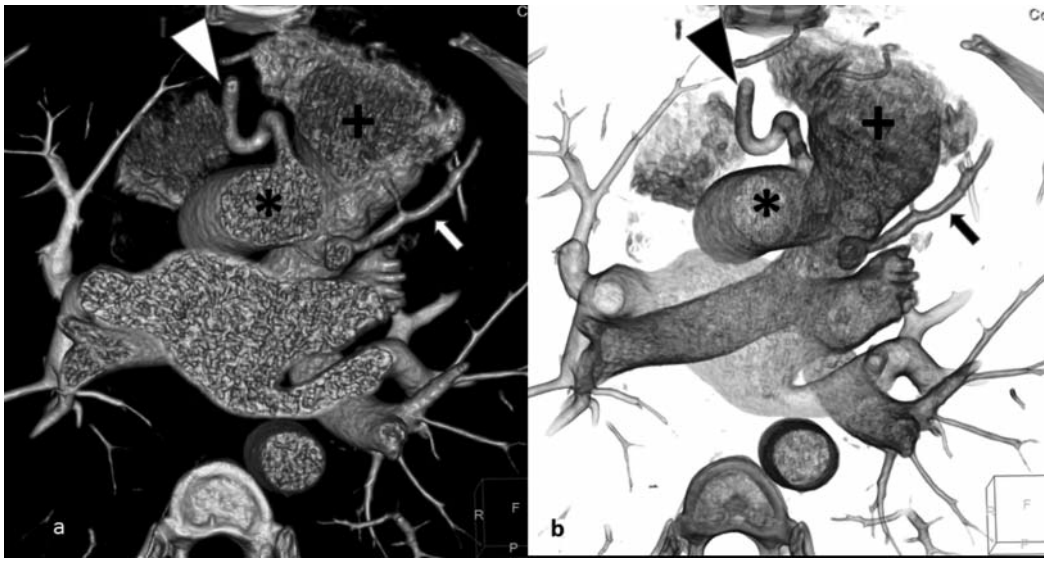

Figure 2. — (a and b) Large arrow heads (black and white) demonstrate the right main coronary artery arising form the ascending aorta (*). The small arrows (black and white) display the anomalous left main coronary artery arising from the inferior pulmonary trunk (+).

Cardiac CT angiography technique

Scanner acquisition

With the patient positioned supine and arms placed above his head, ECG-gated cardiac/coronary CTA was performed using a 256-MDCT scanner. Antero-posterior and lateral scout scans were performed, with a scan range from the apex of the chest to the costophrenic angle. Scan parameters were: detector width 256 × 0.625 mm, pitch 0.2:1, rotation time 0.27 sec, 100 kVp, 200 mA, with z-axis modulation, and scanning time of 2.1 seconds.

Image reconstruction

The following parameters were set: standard image reconstruction of axial images at 0.625 mm slice width, reconstruction interval of 0.5 mm, 180 × 180 mm field of view, and iterative reconstruction technique software (iDose4) with a window width and level of 420 and 65, respectively. The ECG gated scan reconstruction interval with the least motion artifacts was determined by reconstructing a slice at the mid segment of the right coronary artery in 2% steps from 35% to 75% of the R-R interval. For diagnostic interpretation, reconstruction of the CTA images was used, with a time point with the least motion artifact located at the mid segment of the ascending aorta (68%).

Discussion

An anomalous origin of the coronary arteries from the pulmonary artery (PA) is usually an isolated abnormality, it occurs in 0.4% of all patients with heart disease [1]. The most common defect is the anomalous origin of left main coronary artery (LMCA) from the pulmonary artery, known as BWG syndrome [2].

During fetal life the condition can be tolerated due to equal pressure between the PA and the systemic circulation. Shortly after birth, the PA pressure drops and the flow in the LMCA decreases and then reverses leading to a chronic myocardial ischemia and maybe infarction. During this period collateral circulation begins to develop between the RCA and the LMCA; the extent of these collaterals is important in determining the degree of myocardial ischemia, the timing of clinical presentation, and the chances of long term survival [3, 4]. Ninety percent of cases present during

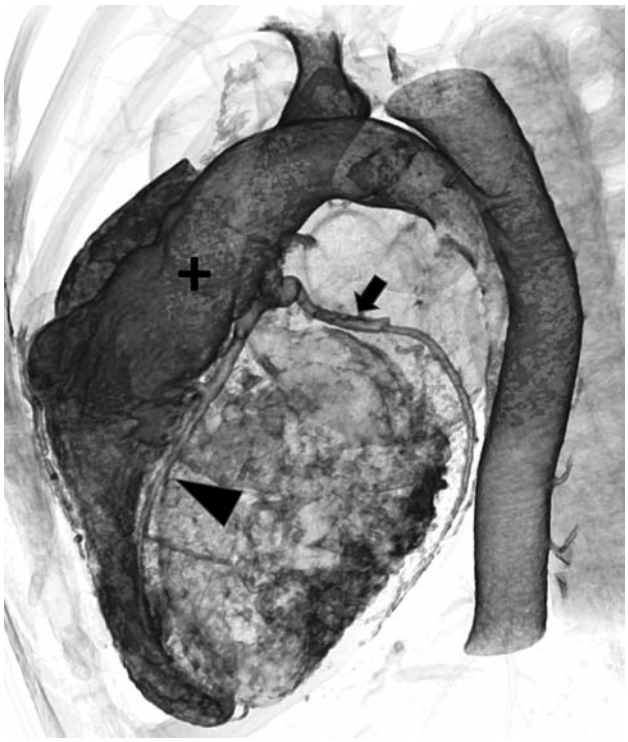

Figure 3. — Demonstrates the left anterior descending coronary artery (large arrow head) and left circumflex coronary artery (small arrow) arising from the inferior pulmonary trunk (+).

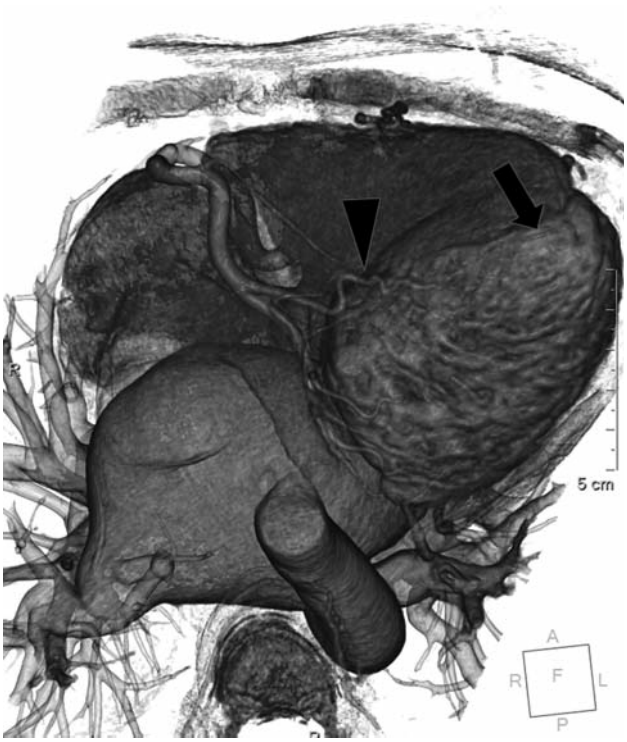

Figure 4. — Demonstrates anastomosis between the right posterior descending coronary artery (large arrow head) and left anterior descending coronary artery (small arrow).

infancy with symptoms of heart failure and mitral regurgitation due to progressive myocardial ischemia, with a bad prognosis and an increased mortality risk during infancy and early childhood [4]. In the adult population it usually presents with a wide range of non-specific symptoms, including syncope, dyspnea on exertion, and angina or cardiac arrhythmias [5]. Very few reported cases were asymptomatic and diagnosed accidentally when following up for other conditions.

Although normal physiologic changes occurring during pregnancy, such as increased oxygen consumption, cardiac output (50%), heart rate, and aortocaval compression in the supine position might increase the burden on the heart [6], yet some uncomplicated cases of anomalous left coronary artery from the pulmonary artery (ALCAPA) in pregnancy have been reported [7].

The anomalous origin of the LMCA from the PA is usually suspected when transthoracic echocardiography shows a dilated RCA arising from the aorta, diastolic blood flow from the LCA into PA, diastolic blood flow from the inferior portion of the inter-ventricular septum to its superior portion, and mitral regurgitation. However these findings are not specific and are shared with other diseases such as Kawasaki's disease and arteriovenous fistula. In order to establish a firm diagnosis cardiac catheterization is used and it shows dilated and tortuous RCA, collaterals, and shunting to the PA [8, 9]. In addition CT angiography is a non-invasive procedure that can help in diagnosis as it can show coronary arteries with a good image quality and high diagnostic accuracy [10].

Many treatment options have been suggested for adults presenting with anomalous origin of the LMCA from the PA yet no optimal surgical technique have been defined. Treatment options include LMCA ligation, re-implantation of the LMCA to its original site in the aorta, baffle creation through the PA (Takeuchi procedure), and a combination of LMCA ligation and coronary artery bypass graft (CABG). Re-implantation of LMCA to the aorta is the first choice of treatment because it restores normal anatomy and circulation; if not possible ligation and CABG are preferred because it also provides a dual coronary flow system [11, 12].

Conclusion

In conclusion, the presented patient is one of the rare cases who survived a deadly disease up until the age of 42 with three pregnancies. A relatively preserved left ventricular systolic function could be explained by having a very large and dominant right coronary artery with adequate retrograde supply to the left system. Again the authors still be-

lieve that the ultimate treatment of this disease is surgery.

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