

## Case Reports

### ANOMALOUS LEFT CORONARY ARTERY ARISING FROM THE PULMONARY ARTERY IN AN ADOLESCENT: NORMALIZATION OF LV DIMENSION AND FUNCTION FOLLOWING REPAIR

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Bland-White-Garland syndrome of anomalous left coronary artery from pulmonary artery is a rare congenital heart disease. It has an incidence of one per 300,000 live births, and represents 0.5% of cases of congenital heart disease.<sup>1</sup> The vast majority of patients present in infancy with a picture of congestive heart failure and angina. Almost all die in the first year of life if untreated. Few reports in the literature describe older individuals with the disease. Our report describes a 12-year-old girl who presented for the first time with exertional dyspnea, and was diagnosed and treated successfully.

#### Case Report

A 12-year-old girl presented with shortness of breath, easy fatigability and episodic nonspecific chest pain of four months' duration. She had been diagnosed earlier in another institution to have idiopathic dilated cardiomyopathy, and had been started on antifailure treatment. Physical examination revealed low active normal growth parameters. Vital signs were within normal limits. The precordium was active on inspection, with clinical evidence of cardiomegaly, and the heart sounds were distant. A grade II/VI faint holosystolic murmur was heard over the apex with no radiation. The liver had normal span and peripheral pulses had low volume.

Chest x-ray showed a cardiothoracic ratio of 0.7, with increased pulmonary vascular markings and prominent hilar vessels. EKG showed sinus rhythm rate at 75 beats/minute and left-axis deviation. *Q*-waves and inverted *T*-waves were apparent in the anterior lateral leads. Left ventricular (LV) strain pattern consistent with left ventricular hypertrophy (LVH) was noted (Figure 1).

Echocardiogram showed atrial situs solitus with intact atrial and ventricular septums. The left ventricular dimensions were significantly increased. The LV had an ejection fraction of 45% and fraction shortening of 20%.



FIGURE 1. A 12-lead ECG showing *Q*-waves and inverted *T*-waves in the anteriolateral leads, as well as left ventricular strain pattern consistent with LVH.

The right atrium (RA) was prominent. The proximal left coronary artery (LCA) was not seen. By color-Doppler mapping, there was a diastolic run-off in the main pulmonary artery (MPA) and mild mitral regurgitation. Coronary artery fistula vs. anomalous LCA from the MPA was suspected.

Thallium treadmill exercise test showed fair exercise tolerance, however, was inconclusive for ischemia. A positron emission tomography (PET) scan showed viable myocardium in the circumflex and left anterior descending (LAD) territories.

Cardiac catheterization and angiography were performed. Oxygen saturation data showed a step-up in the main pulmonary artery to 78% from 55% in the superior vena cava (SVC), resulting in a calculated pulmonary-to-systemic flow ratio of about 2. Right ventricular pressures were normal, however, the left ventricular end diastolic pressure was slightly elevated to 15 mm Hg.

Selected right coronary artery angiogram revealed a slightly dilated vessel with extensive collateral circulation into the distal circumflex and LAD arteries, which were filled in a retrograde manner. Eventually the contrast was seen filling the MPA through the communication with the LCA (Figure 2). The ascending aortogram failed to show any communication between the coronary cusps and the LCA. LV angiogram showed akinetic apex and hypokinesia of the antero-lateral LV wall.

After establishing the diagnosis, the patient underwent surgical repair, where the LCA was mobilized off the

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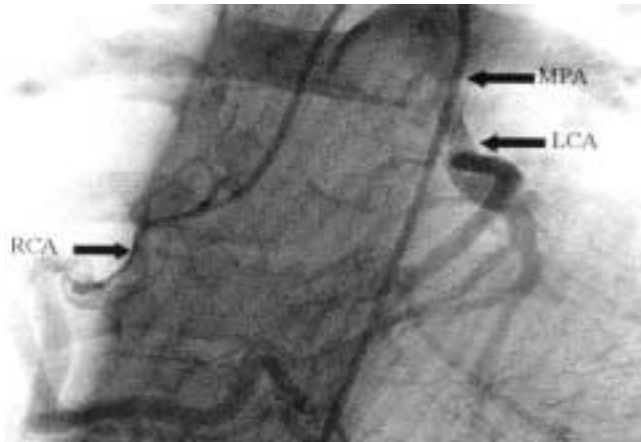


FIGURE 2. Right coronary artery angiogram showing a dilated vessel with extensive collateral communication with the left coronary artery originating from the main pulmonary artery.

anterior pulmonary sinus with a generous cuff of tissue. However, due to the wide gap between the aorta and the position of the severed LCA, a tunnel was constructed. A tongue of tissue from the pulmonary artery and another from the aorta were used to construct the tunnel and establish a two-coronary system. The aortotomy was closed primarily, and the pulmonary artery was patched using autologous pericardium.

On her latest visit one year following the procedure, the patient was totally asymptomatic. Echocardiography showed remarkable improvement in the LV function, with an ejection fraction of 67% and the fractional shortening of 32%, however, the apex remained akinetic.

### Discussion

Bland-White-Garland syndrome is a rare congenital heart disease. If left untreated, the vast majority of infants diagnosed with this disease will not survive beyond their first birthday. The patient might be initially asymptomatic due to the elevated pulmonary vascular resistance and pressure, hence the perfusion of the anomalous coronary artery may remain adequate. Most patients, however, will develop symptoms as the pulmonary vascular resistance and pressure drop. Angina-like episodes during this stage are frequently misinterpreted as colics. If the patient survives, collateral circulation might develop from the right coronary artery to the left coronary artery. Eventually, a "coronary steal" phenomenon will develop, as the blood will preferentially flow from the RCA into the MPA

bypassing the myocardium. Ischemia, especially with exertion, is described at this stage. The diagnosis of congestive cardiomyopathy in our patient is most probably due to the "steal phenomenon" resulting in an increase in the pulmonary-to-systemic flow ratio.

Several surgical techniques have been advocated for the repair of this anomaly. Those techniques were reviewed by Sabiston et al.,<sup>3</sup> and Neufeld and Schneeweiss.<sup>4</sup> Those procedures aimed at improving myocardial perfusion or oxygenation, collateral circulation, or increasing myocardial perfusion pressure.

Currently, most centers consider re-establishment of the two-coronary artery system by aortic reimplantation as the procedure of choice.<sup>5-8</sup> Although the conclusive superiority of establishing the two-coronary artery system has not yet been demonstrated, it is intuitively more attractive and physiological.

Our patient underwent successful direct aortic reimplantation of the left coronary artery with very good short-term outcome. This favorable outcome was evident by clinical improvement as well as electrocardiographic and echocardiographic parameters. Left ventricular systolic function normalized after four months following surgery. The improvement of the patient's dilated cardiomyopathy might be in part due to the elimination of her left-to-right shunt, however, improved myocardial perfusion might have played a role as well.

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