

Case 5/2012 – Six-Year-Old Child with Anomalous Origin of Left Coronary Artery and Mitral Regurgitation

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Clinical data

Tiredness, noticed since three days after birth, persisted during the first year, while receiving digoxin and captopril. Thereafter, the child remained asymptomatic and had full physical capacity, while taking enalapril (5 mg/day).

Physical Examination: good general status, eupneic, acyanotic, normal pulses in all four limbs. Weight: 20 Kg; BP: 100/70 mmHg; HR: 80 bpm; O₂ Saturation: 96%. Aorta not palpable at the suprasternal notch.

In the precordium, the *ictus cordis* was palpable between the 4th and 5th intercostal spaces, in the hemiclavicular line, by two digital pulps. Heart sounds were normal and slight systolic murmur was heard, + intensity, in the mitral area without radiation. The liver was not palpable and lungs were clear.

Complementary examinations:

Electrocardiogram

Showed sinus rhythm and signs of left ventricular overload with Sokoloff index (S of V2 and R of V6) of 57 mm. T wave was negative in the anterolateral wall, AP: +50°, AQRS: +20°, AT: +100° (Figure 1).

Chest X-ray

Showed enlarged heart silhouette at the expense of the left ventricular arch and left atrium. A rectified middle arch was observed and the pulmonary vascular network was slightly increased (Figure 1).

Echocardiogram

Shows dilated left heart cavities, moderate regurgitation of the thickened mitral valve and prolapse of the anterior valve, hyperrefringence of the left papillary muscle, dilated right coronary artery and anomalous origin of the left coronary

Keywords

Heart defects, congenital; mitral valve insufficiency; coronary vessel anomalies; thoracotomy.

artery of the pulmonary trunk in the left anterior sinus of the pulmonary valve. Heart measurements were as follows: RV = 16, LV = 52, LA = 32, Ao = 19, Tricuspid Annulus = 21, Mitral Annulus = 28, RPA=LPA = 11, LVEF = 61%, right coronary artery = 5 mm, left coronary artery = 4 mm (Figure 2).

Cardiac catheterism

Showed at angiography the same elements with abundant collateral circulation between the two coronary arteries and clear flow direction from the left coronary artery to the pulmonary trunk (Figure 2).

Clinical diagnosis

Anomalous origin of left coronary artery from the pulmonary trunk with abundant collateral circulation between the coronary arteries and preserved ventricular function, but with moderate mitral valve regurgitation.

Clinical rationale

The clinical elements were compatible with the diagnosis of acyanogenic congenital heart disease with no dynamic effect, considering good physical tolerance and no signs of heart failure. Cardiac auscultation supported the diagnosis of mitral regurgitation of moderate outcome, in agreement with the radiographic imaging tests and electrocardiograms. Echocardiography and coronary angiography established the accurate diagnosis of the anomaly. It was difficult to characterize the cause of the mitral regurgitation due to the normal ventricular function in the presence of coronary anomaly. Hyperrefringence of the papillary muscles, an expression of prior ischemia, suggested the ischemic cause, notwithstanding the preserved ventricular function. The association of mitral valve prolapse, regardless of the basal lesion, was also suggested.

Differential diagnosis

Other congenital heart diseases that are associated with left ventricular volume overload can also manifest by causing mitral regurgitation due to mitral annulus dilatation. Most notable are the aortopulmonary window, persistent ductus arteriosus and other arteriovenous fistulas. However, these anomalies would show other clinical manifestations, especially continuous murmur in the precordium or another organic site.

Conduct

Enlargement of the two coronary arteries, particularly of the right one, was found through median thoracotomy. The

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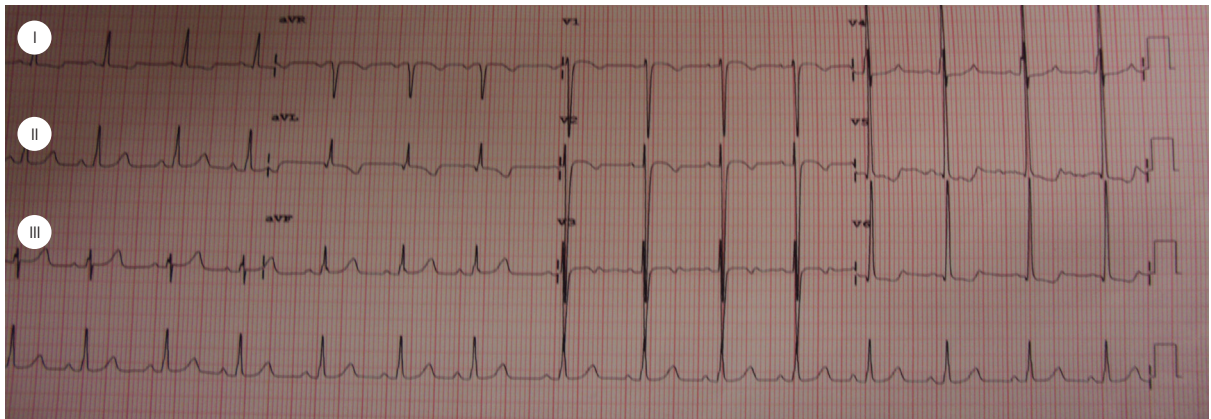
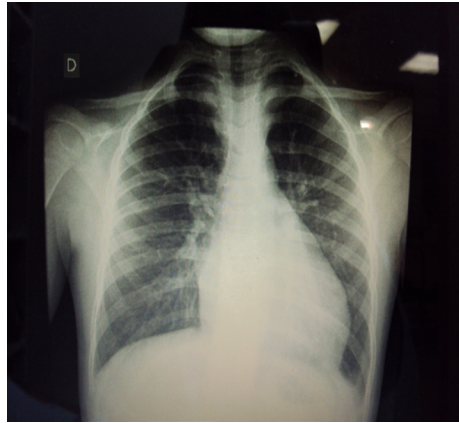
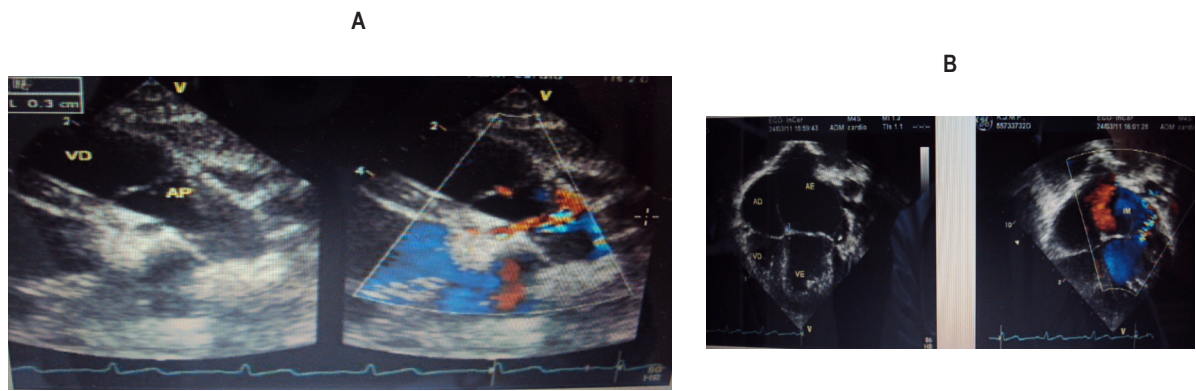


Figure 1 – Chest X-ray highlights enlarged cardiac area due to prominent left ventricular arch, rectified medium arch and somewhat increased pulmonary vascular network in the pulmonary hila. The electrocardiogram highlights signs of left ventricular overload and anterolateral ventricular repolarization alterations.



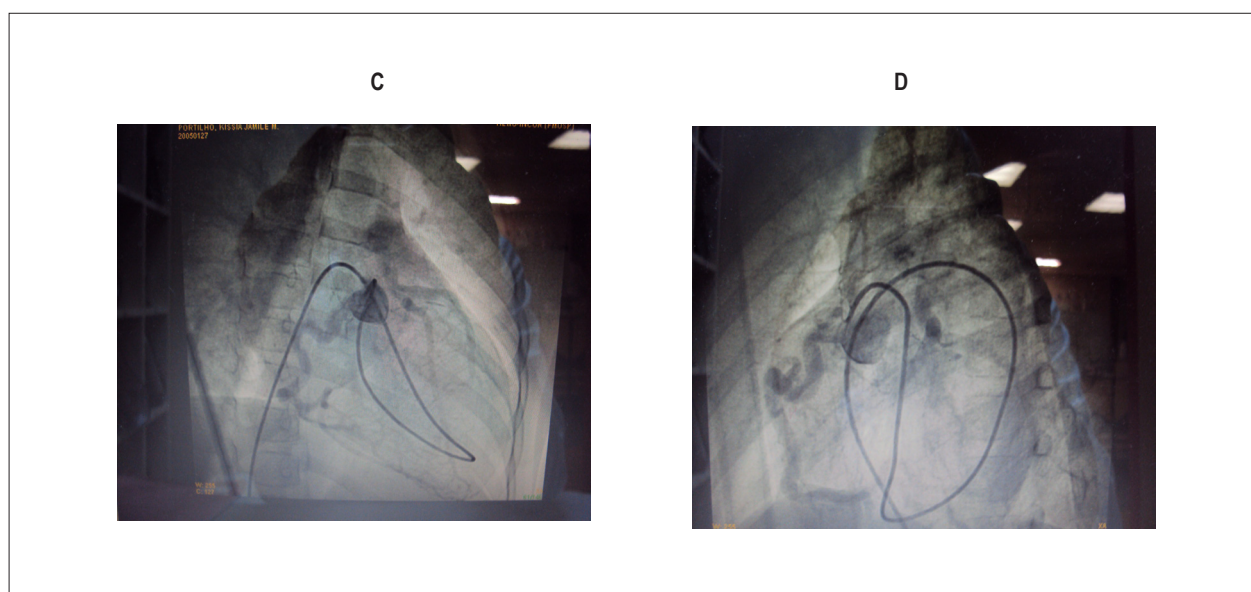


Figure 2 – The echocardiogram shows, in addition to the anomalous origin of left coronary artery from the pulmonary trunk at subcostal view and in color in A, enlarged left cavities in the apical four-chamber view, due to mitral prolapse and regurgitation in B. The angiography shows a dilated right coronary artery, abundant collateral circulation with filling of the left coronary artery showing blood flow to the pulmonary trunk, in C (right anterior oblique view) and D (left anterior oblique view).

anomalous left coronary artery was transported directly from the left posterior sinus of the pulmonary artery to the lateral and posterior wall of the aorta. There was mitral valve prolapse in the anterior valve and annulus dilatation was reduced by posterior plication. The postoperative course was uneventful.

Comments

The diagnosis of anomalous origin of left coronary artery is hindered when the patient shows no signs of myocardial

ischemia in the presence of sufficient collateral circulation between the coronary arteries, thus preserving the arterial blood flow to the myocardium. In this situation, the diagnosis of the anomaly becomes fortuitous due to another factor that precipitates the performance of diagnostic exams. Even without ischemia, the correction of anomalous left coronary artery follows the principle of obtaining coronary circulation fed by two arterial sources, with greater uniformity of myocardial blood flow, as well as preventing a more severe atherosclerotic coronary accident in the long-term evolution at adult age.