Anomalous origin of the left coronary artery from the pulmonary artery in an adult pregnant patient: surgical and percutaneous myocardial revascularization

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An anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) was diagnosed in a 31-year-old woman complaining of angina at 10 weeks of gestation. After termination of pregnancy, the patient underwent surgical repair with ligation of the left coronary artery at the ostium, and a single bypass graft with a left internal thoracic artery to the left anterior descending coronary artery. Angiography, performed at 6 months of follow-up, showed stenosis of the distal anastomosis of the graft that was treated with angioplasty and deployment of a paclitaxel-eluting stent. After 9 months the patient was symptom-free and angiography excluded in-stent restenosis.

Experience with the surgical repair of the anomaly is limited by the rarity of this condition; in particular, it has never been reported as a first diagnosis during pregnancy. In the discussion, we analyze the efficacy and limits of the different current strategies for the surgical repair of this rare, but potentially life-threatening, congenital coronary anomaly.

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Introduction

Congenital coronary artery malformations are rare anomalies that may nevertheless be life-threatening. It is estimated that they constitute the underlying cause of sudden cardiac death in up to 35% of cases in young people[1].

The incidence of an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is 1 per 300 000 live births[1]. This condition is characterized by decreased left coronary perfusion and hence a possibly impaired ventricular function. In case of the onset of heart failure, the prognosis generally depends on the promptness of surgical repair and on the degree of left ventricular dysfunction[1-3]. Death generally ensues within the first year of life. Occasional cases diagnosed in adulthood, however, have been described[1]. We report a case of an adult woman with ALCAPA and a preserved left ventricular function who first presented during pregnancy.

Case report

A 31-year-old woman, with history of mitral valve prolapse presented, at the 10th week of gestation, with recent-onset retrosternal effort chest pressure and discomfort, suggestive of angina. She had no known risk factors for coronary artery disease. Physical examination revealed an apical mid-systolic murmur but was otherwise unremarkable. ECG showed complete right bundle branch block (Fig. 1A). Somewhat less typical symptoms in the past had been ascribed to mitral valve prolapse. In view of the persistence of chest pain and pressure, an exercise stress test was performed. The stress test was positive for inducible ischemia with ST-segment depression in the precordial leads (V_2 to V_6) and angina (Fig. 1B).

An echocardiogram was suggestive of anterolateral wall hypokinesis, even though the ejection fraction was normal (57%). The proximal segments of the coronary arteries appeared enlarged with the left main originating from the posterior wall of the main trunk of the pulmonary artery. Color Doppler showed reverse flow in the left coronary artery to the pulmonary artery.

Coronary angiography documented ectasia and tortuosity of the right coronary artery with an extensive collateral circulation to a left coronary artery originating from the pulmonary artery trunk (Fig. 2).

Because of stress-induced ischemia at a very low workload, it was recommended

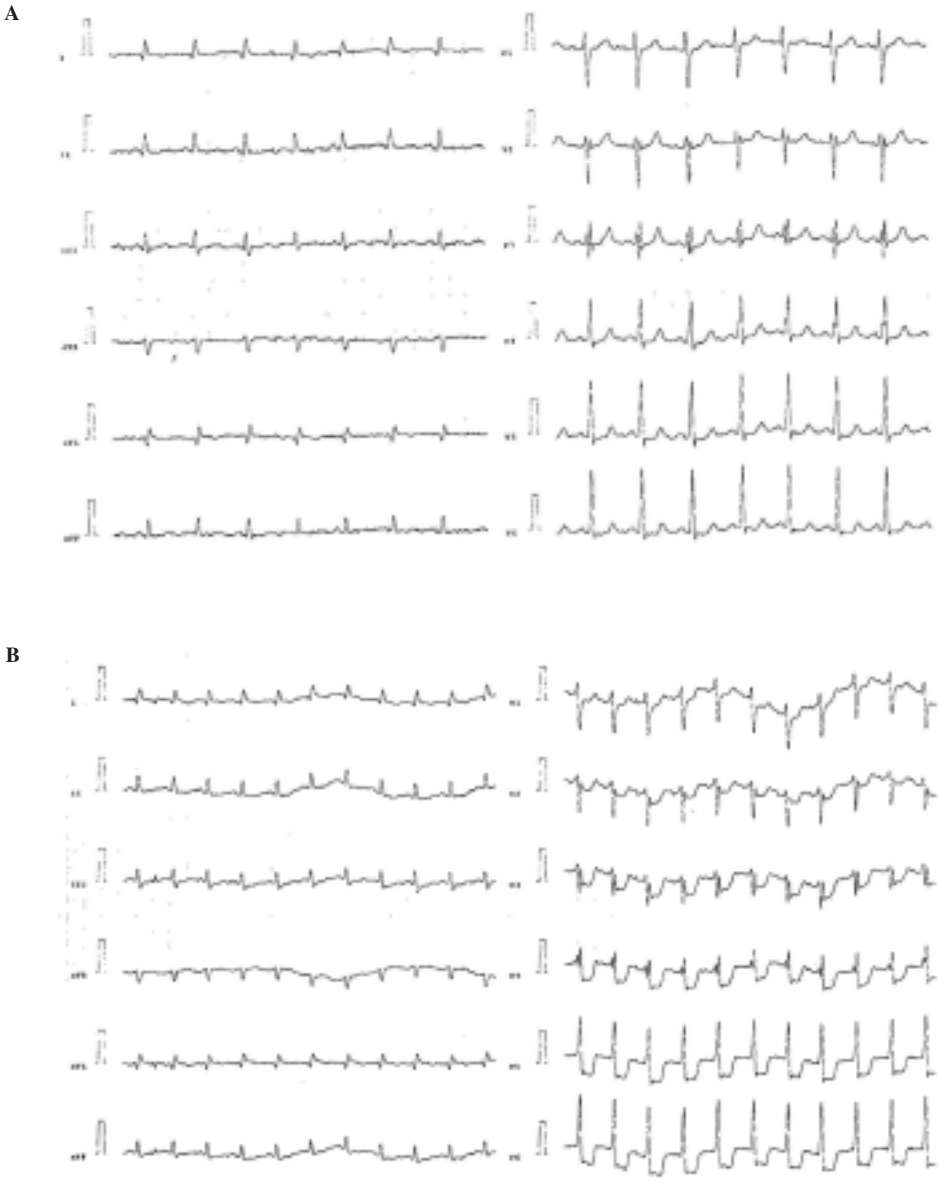

Figure 1. A: *rest ECG;* B: *stress ECG.*

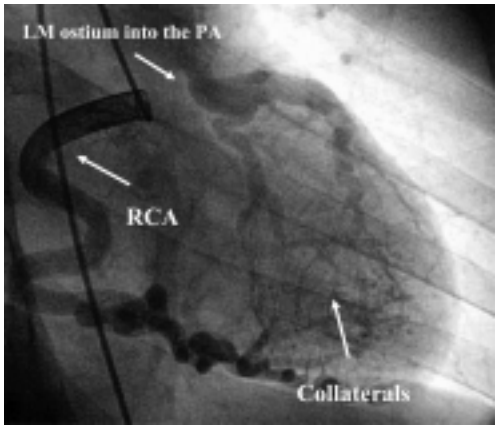

Figure 2. *Right coronary angiography showing an extensive collateral circulation to the left coronary artery originating from the pulmonary trunk. LM = left main; PA = pulmonary artery; RCA = right coronary artery.*

that the patient end her gestation. Following termination of pregnancy, the patient underwent surgical repair with ligation of the left coronary at the ostium and a single bypass graft with a left internal thoracic artery to the left anterior descending coronary artery. Myocardial protection was obtained by cardioplegia and left ventricular support was not required. The postoperative course was uneventful and the patient remained asymptomatic.

A stress test performed after 3 months was again positive. Angiography showed stenosis at the perianastomotic site of the graft and diffuse dysplasia of the left coronary artery (Figs. 3 and 4). The patient underwent angioplasty of the anastomosis of the graft with deployment of a paclitaxel-eluting stent (TAXUS stent, Boston Scientific Corporation) with optimal immediate angiographic results (Fig. 5).

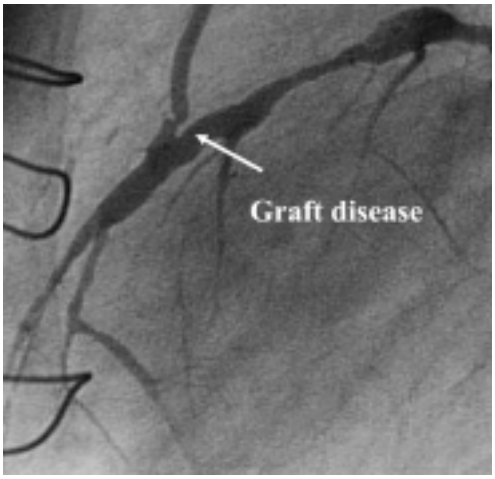

Figure 3. *Bypass angiography in the lateral view showing distal graft disease and diffuse disease of the left anterior descending coronary artery.*

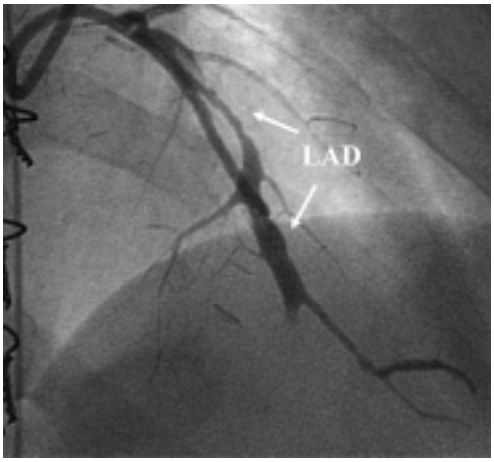

Figure 4. *Left anterior descending coronary artery (LAD) as seen at bypass angiography in the cranial anterior view showing diffuse disease of the artery.*

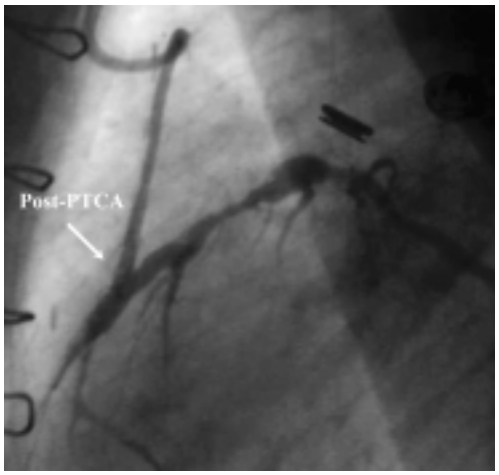

Figure 5. *Bypass angiography in the lateral view showing the post-angioplasty (PTCA) outcome.*

At 9-month clinical follow-up, the patient was still asymptomatic. A repeat stress test was performed and was found to be normal at 13 METS and 95% of the predicted maximum heart rate, with no symptoms and no ECG changes. Angiography revealed patency of the distal bypass anastomosis (Fig. 6).

Discussion

The Bland-White-Garland syndrome (ALCAPA) represents about 0.4% of congenital cardiac anomalies and is the most common congenital coronary malformation[1]. This malformation is the result of displacement in the process of partition of the truncus arteriosus, around which the proximal coronary network has been formed. In this anomaly, during fetal life, the left coronary blood flow from the pulmonary trunk is antegrade, since vascular resistance in the pulmonary circulation is higher than in the systemic circulation. After birth, with the fall in pulmonary artery resistance, the direction of blood flow in the left coronary artery reverses and left ventricular perfusion depends on the degree of collateral circulation from the right coronary artery[4,5]. In these conditions survival is allowed by an extensive collateral circulation in only 10-15% of cases[6].

Most patients develop severe myocardial ischemia with signs of congestive heart failure. Children usually complain of angina and dyspnea and occasionally syncope on crying or eating. Older children and adults generally develop dilated cardiomyopathy with functional mitral regurgitation, malignant arrhythmias or sudden death (80-90% mortality estimated at a mean age of 35 years)[1].

Our case is very unusual because the patient presented during pregnancy. It is well known that during pregnancy there is a progressive increase in cardiac output and heart rate[7]. In our patient, the mild increase in oxygen consumption normally occurring in the first

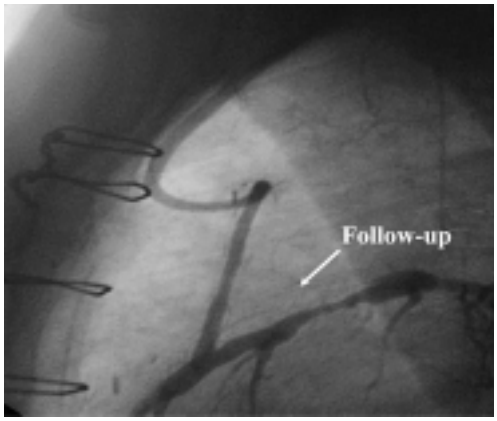

Figure 6. *Nine-month angiographic follow-up.*

trimester, may have been sufficient for the development of chest pain on exercise. Because of the risk of ischemia and cardiac failure by the end of pregnancy and delivery, we strongly recommended that the patient terminate her pregnancy.

As described in the literature, the diagnosis of the anomaly was made at echocardiography. Medical therapy is considered only palliative and is indicated to treat myocardial infarction, left ventricular dysfunction or arrhythmias.

Therapy consists of surgical repair of the anomaly. Even though the extreme rarity of the disease limits the experience in this field, different surgical approaches have been described:

• simple ligation of the origin of left coronary artery, with restoration of a single coronary artery system[8];

• direct reimplantation of the anomalous left coronary ostium into the aorta with excision of a large button of the pulmonary artery[2];

• the Takeuchi technique, initially proposed for cases in which direct reimplantation was not feasible (i.e. ostial stenosis, lack of coronary artery length), consists of a coronary tunnel inside the pulmonary artery constructed using a baffle of pulmonary artery wall, starting from an aortopulmonary window into the left coronary ostium[9];

• the interposition of a segment of the free subclavian artery between the aorta and the coronary ostium described by Arciniegas, as a variant of the Takeuchi operation[1,3].

• bypass grafting[3,6];

• orthotopic heart transplantation. This technique has been described in case of persistent myocardial dilation after initial surgical myocardial revascularization[10].

Having restored the blood flow, high rates of improvement in the degree of mitral regurgitation have been described postoperatively. For this reason, concomitant mitral valve surgical repair still remains controversial[1-3].

Nevertheless, persistent severe mitral regurgitation is one of the main reasons for reintervention[1,3]. Generally, the current policy is to perform mitral valve repair only in patients with truly severe mitral regurgitation[1].

The late prognosis after successful surgical repair has not been well defined. However, the most important predictor of long-term survival is probably the degree of residual left ventricular systolic function after coronary flow restoration. In this scenario, better results have been achieved by restoring a dual coronary circulation.

Because of the favorable preoperative characteristics (normal left ventricular systolic function and the absence of significant mitral regurgitation) a single bypass graft with an arterial conduit seemed to be a good option in our case. This surgical technique had been previously described in a few case reports[1,11] and seemed to be one of the best strategies for adult patients with ALCAPA. Nevertheless, in our case graft failure unfortunately occurred. This is considered a very rare event with arterial conduits.

Retrospectively, more complete surgical revascularization with multiple conduits may have been preferable in our patient who presented with an associated extensive dysplasia of the left coronary tree. Concern should arise on the need of a more accurate preoperative diagnosis of possible associated coronary anomalies in such patients, in whom selective left coronary angiography cannot be performed until after surgical repair. Preoperative multislice coronary computed tomography scanning may be helpful for risk stratification[12].

Following the diagnosis of bypass failure we opted for percutaneous revascularization. Angioplasty and stenting of an arterial bypass graft at the anastomotic/perianastomotic site is usually limited by a 20 to 30% restenosis rate[13,14]. We deployed a drug-eluting (paclitaxel) stent to lower the risk of recurrence[15].

In summary, ALCAPA is a rare but potentially lethal congenital coronary anomaly. Pregnancy may exacerbate symptoms. We describe an unusual case of angioplasty and drug-eluting stenting at the anastomotic site after unsuccessful surgical revascularization with a single bypass graft. A review of the literature suggests that left coronary artery bypass grafting with multiple arterial conduits may be the treatment of choice for this rare subset of patients.

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