*Case Report*

# A Surgical Report of Bland White Garland Syndrome

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**We report a 22-year-old mother of 2 who presented to us with exertional chest pain for 3 years. On coronary angiography she was diagnosed to be suffering from Bland White Garland syndrome (BWGS). She had a giant and grossly tortuous right coronary artery (RCA) forming collaterals with the left coronary artery (LCA), which was draining into the pulmonary artery (PA). Surprisingly, she had no evidence of mitral regurgitation on echocardiography, and she had a preserved left ventricular systolic function. She underwent Takeuchi's repair with uneventful recovery. Postoperative CT angiography revealed adequate reimplantation of the left main coronary artery to the aorta with patent tunnel. On a 6-month follow-up, she is asymptomatic and has an optimal flow through the tunnel to the LCA. (Ann Thorac Cardiovasc Surg 2009; 15: 53–57)**

**Key words:** anomalous left coronary artery from the main pulmonary artery, Bland White Garland syndrome, coronary artery anomaly, Takeuchi's repair, congenital heart failure

## Introduction

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA), also known as Bland White Garland syndrome (BWGS), is a rare congenital anomaly. It accounts for 1 in 300,000 live births (0.25% to 0.5%). It usually occurs as an isolated lesion.[1,2)]

In infants, it presents as failure to thrive, profuse sweating, dyspnoea, pallor, and atypical chest pain on eating or crying.[3,4)] Presentation in adults is extremely rare with symptoms of ischemia, congestive cardiac failure, mitral regurgitation, and malignant arrhythmias leading to sudden death.[4,5)] Surgical correction is the "gold standard" for the BWGS therapy because surgically uncorrected cases have 80%–90% mortality at a mean age of 35 years.[4,6)] Among the surgical approaches suggested are ligation of anomalous left coronary artery (LCA), various forms of bypass grafts from the aorta, subclavian to LCA anastomosis, transpulmonary baffling or Takeuchi's procedure, and direct reimplantation of the coronary artery into the aorta.[5)] Coronary transfer is the recommended technique of choice.[5)]

## Case Report

A 22-year-old mother of 2 had a history of recurrent chest pain for 3 years, which progressed to angina on ordinary physical activity, when she was diagnosed with BWGS. The pain radiated to the left arm and was associated with palpitations, sweating, and shortness of breath. This was relieved with rest. The first time she had symptoms was when she was pregnant during her first trimester 3 years ago. It was attributed to acid peptic disease (APD) and anemia. After the birth of her

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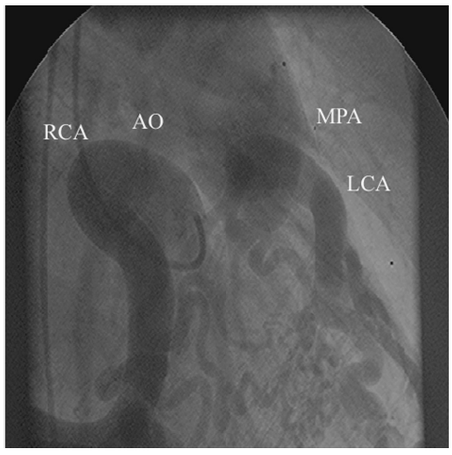


**Fig. 1.** Selective injection of RCA by Judkin's right catheter, showing a grossly dilated and tortuous RCA giving extensive collaterals to LCA, which had anomalous origin from the MPA.
RCA, right coronary artery; AO, aorta; MPA, main pulmonary artery; LCA, left coronary artery.

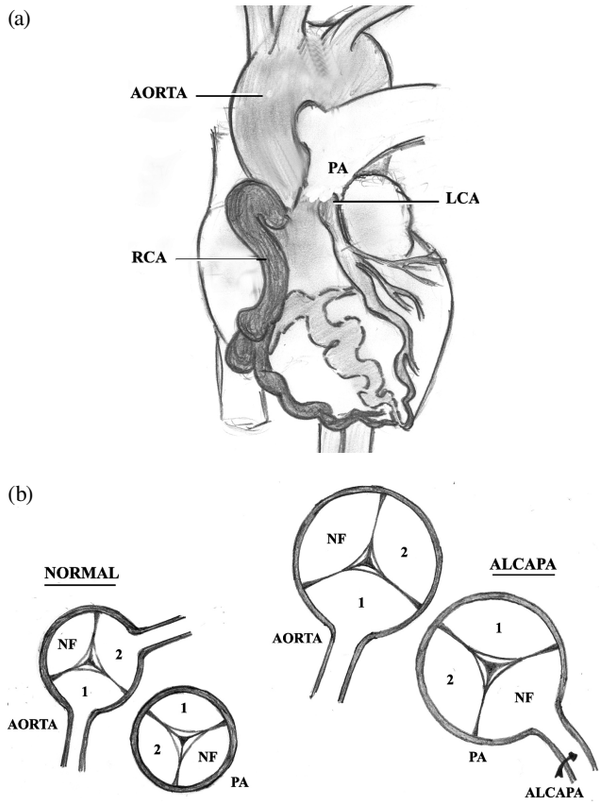


**Fig. 2.**
**a**: Diagram showing LCA arising from PA, dilated tortuous RCA giving collaterals to LCA.
RCA, right coronary artery; PA, pulmonary artery; LCA, left coronary artery.
**b**: Normal anatomy shown on left panel of figure; the right panel shows anomalous LCA arising from the nonfacing sinus of PA.
NF, nonfacing sinus; PA, pulmonary artery; ALCAPA, anomalous left coronary artery arising from the pulmonary artery; LCA, left coronary artery.

baby, she was asymptomatic. These symptoms reoccurred during her second pregnancy, and since than the severity and intensity has increased. She was treated for APD once again, but with no relief. After a delay of one year she came to our institute for diagnosis of chest pain.

Examination of the cardiovascular system revealed a heart rate of 78 per minute, blood pressure of 110/70 mm Hg, and a respiratory rate of 18 per minute. There was a grade 3/6 continuous murmur at the left upper sternal border with no respiratory variation. Her body mass index was 18.5 kg/m$^2$. Transthoracic and transesophageal echocardiography was done, which revealed a dilated right coronary artery (RCA). The origin of LCA could not be visualized, but there was turbulence in the pulmonary artery (PA) with continuous flow, features suggestive of BWGS. She underwent cardiac catheterization, which revealed LCA not arising from the left coronary sinus. A selective injection of RCA showed a grossly dilated and tortuous RCA giving extensive collaterals to LCA, which had anomalous origin from the main PA (MPA). By this injection, the MPA and its branches were opacified (Fig. 1). Anomalous LCA was seen to be arising from the nonfacing sinus of PA (Fig. 2). She was then referred for surgery. Median sternotomy was done, and pericardiotomy revealed tortuous left and right coronary arteries (Fig. 3). The left main stem (LMS) identified attached to the lateral side (nonfacing sinus) of the PA.

After a median sternotomy, extracorporeal circulation was established in a routine manner with moderate hypothermia. Crystalloid cardioplegia (St. Thomas Hospital solution) was infused into the ascending aorta after cross-clamping. The coronary anatomy was assessed, and we found that adequate mobilization of anomalous LCA was not possible. Therefore we were unable to implant it as a button on the aorta. So we proceeded to do a Takeuchi's repair by making an intrapulmonary baffle using the anterior wall of PA. This was than anastomosed with the aorta. The resulting defect in the PA was closed with an autologous pericar-

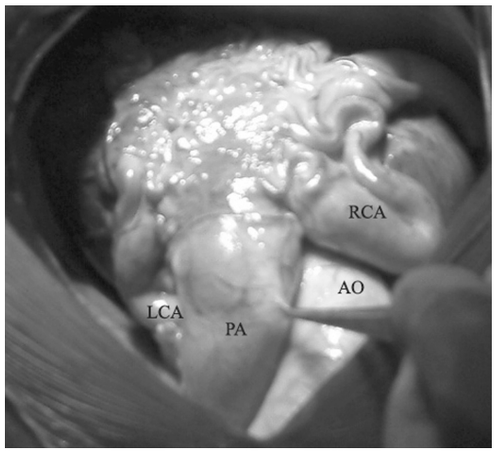


**Fig. 3.** Tortuous right and left coronary arteries, the LCA seen arising from the PA. LCA, left coronary artery; PA, pulmonary artery; RCA, right coronary artery; AO, aorta.

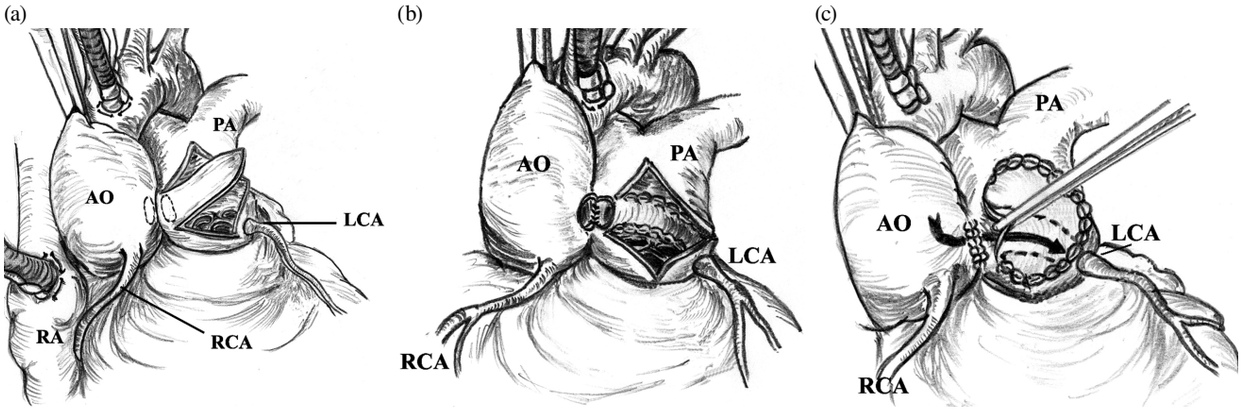


**Fig. 4.**

**a:** A baffle of PA tissue is being made between the AO and anomalous LCA by means of a flap of PA's anterior wall. PA, pulmonary artery; AO, aorta; LCA, left coronary artery; RA, right atrium; RCA, right coronary artery.

**b,c:** Showing the intrapulmonary baffle constructed using the anterior wall of the PA, which is then reconstructed with an autologous pericardial patch.

AO, aorta; PA, pulmonary artery; RCA, right coronary artery; LCA, left coronary artery.

dial patch. The patient was rewarmed and weaned off cardiopulmonary bypass uneventfully (Fig. 4). The postoperative recovery was also uneventful. A postoperative CT angiography was performed, which revealed adequate reimplantation of the LMS to the aorta (Fig. 5). The patient has been asymptomatic since the operation with no complaints on follow-up. An echo at 3 months showed normal PA and flow of blood from aorta to the LCA via the repair. There was no posttunnel dilatation. At the 6-month follow-up, the patient was asymptomatic, and coronary angiography was performed to visualize the integrity of the tunnel. It revealed optimal flow through the tunnel to the LCA with no complications as a result of Takeuchi's repair (Fig. 6).

## Discussion

Among congenital coronary artery anomalies, the ALCAPA or BWGS is the most common.[7)]

On the basis of onset of presentation, BWGS has two main types, infantile and adult.

The infantile type has an early onset of myocardial ischemia, left ventricular dysfunction and dilatation, mitral regurgitation from papillary muscle ischemia, and dilatation of the annulus. An infantile type of circulation has a lack of collateral development.[4,6)] Without

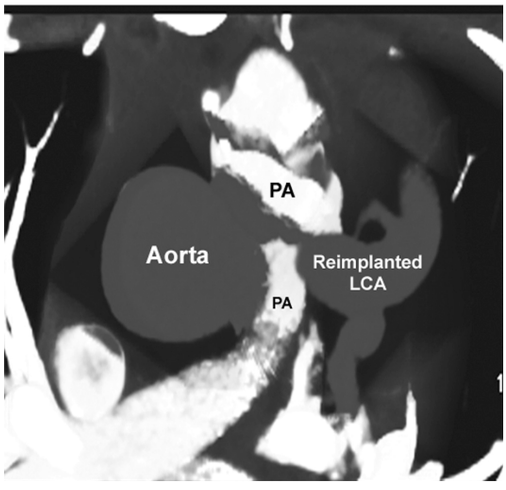


**Fig. 5.** CT angiography image showing a reimplantation of the LCA to the aorta.
PA, pulmonary artery; LCA, left coronary artery.

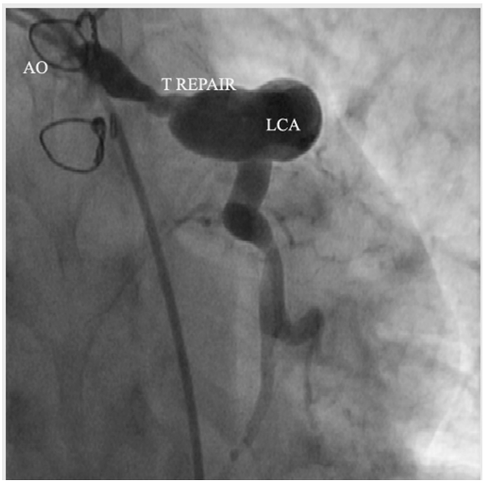


**Fig. 6.** Injection in the left coronary system showing optimal flow through the tunnel (formed by Takeuchi's repair) to the LCA on a 6-month follow-up coronary angiography.
AO, aorta; T REPAIR, Takeuchi's repair; LCA, left coronary artery.

surgical correction, death can occur within the early weeks after birth.[4,6] The adult type is found in only 10%–15% of patients. In this subgroup, survival is due to the large dominant RCA with extensive intercoronary collaterals and a restrictive opening between the anomalous LCA and PA.[4,6]

Selzman et al.[8] have reported a young female who presented with flulike symptoms, though she had lived asymptomatically 20 years; however, she denied any symptoms of congestive heart failure.[8] The absence of mitral pathology and abnormal left ventricular function in their patient was due to well-developed collaterals with a developed left to right shunt.[8] Our young female patient had had an asymptomatic infancy and childhood. She first started having symptoms during pregnancy. In spite of her symptoms, though, she had no evidence of mitral regurgitation, and she had a preserved left ventricular systolic function on echocardiography. On the contrary, most adult patients of BWGS present with symptoms of severely impaired left ventricular function and mitral regurgitation. Damage to the left ventricle results from myocardial ischemia leading to endocardial and subendocardial fibrosis, damage to the papillary muscles, patchy myocardial necrosis, and dilatation of the ventricle.[9,10] All these subsequently lead to mitral regurgitation.[9,10] In our patient, probably the excellent collateral formation between the RCA and the left coronary circulation prevented the occurrence of left ventricular dysfunction and mitral regurgitation.

Surgical correction is the treatment of choice once a diagnosis of BWGS has been made because medical management is associated with poor survival and a high mortality ranging from 45%–100%.[4] Surgical intervention is therefore recommended at the time of diagnosis.[11]

In infants, a direct reimplantation of anomalous LCA into the aorta is frequently done, followed in rare cases by Takeuchi's repair. In adults, both direct reimplantation and Takeuchi's repair may be technically more difficult; therefore simpler but equally effective procedures are used. These include ligation of the LCA from the PA or ligation of the LCA from the PA and coronary artery bypass grafting, using the internal thoracic artery or the saphenous vein graft.[11]

Direct reimplantation with a reported mortality of 0%–16% is the most frequently adopted surgical technique. Lange et al.,[5] in their study of 56 patients of BWGS, confirmed excellent results with this technique.[5] The distance between the anomalous LCA and the aorta predicts the choice of surgical procedure because a direct implantation is performed when the distance between the two arteries permits an easy transfer of the anomalous LCA to the aorta. However, if LCA originates from an extremely lateral aspect of the PA, or when the distance does not permit, an easy transfer of Takeuchi's repair is preferred.[12] In our patient, Takeuchi's repair was done because the anomalous LCA ostium was located laterally and close to the

pulmonary valve commissure, and excision with mobilization of the coronary artery would not have generated sufficient arterial wall to support the coronary transfer. Thus a direct implantation of LMS was not performed, even though it would have been an easier operation in comparison to Takeuchi's repair with better results in an adult. Furthermore, bypass grafting to the left anterior descending artery (LAD) by Saphenous vein graft was not done, keeping in view the young age of the patient and the high subsequent occlusion of these grafts over long-term follow-up, subsequently needing a redo operation. The large tortuous vessels would further preclude the adequate long-term outcome of this procedure.

## Conclusions

We conclude that we can safely recommend Takeuchi's repair in adult patients with BWGS without left ventricular dysfunction and mitral regurgitation.

## Acknowledgment

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