

# Repair of Bland-White-Garland Syndrome via a Modified Technique

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*Surgically repairing Bland-White-Garland syndrome (anomalous origin of the left coronary artery from the pulmonary artery) is a challenge if there are variations in the origin of the anomalous artery. We report the successful repair of this congenital abnormality in a 19-year-old woman who presented with an acute anterior myocardial infarction. The anomalous artery originated from the anterior-facing sinus of the pulmonary artery, which precluded typical repair by direct reimplantation or fashioning an intrapulmonary tunnel. We created an extrapulmonary tunnel, using a strip of pulmonary artery anteriorly and an aortic flap posteriorly. Three years postoperatively, the anastomosis was patent and the patient was asymptomatic. Our modified technique might serve as an alternative method during similar surgical circumstances. (Tex Heart Inst J 2014;41(1):48-50)*

**B**land-White-Garland syndrome, or anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), constitutes only 0.5% of congenital cardiac conditions.<sup>1</sup> Without appropriate surgical correction, the condition has a mortality rate as high as 90% in the first year of life.<sup>2</sup> Symptomatic neonates with ALCAPA typically present either with acute myocardial ischemia that leads to systolic dysfunction and shock, or with congestive heart failure caused by poor left ventricular (LV) function and mitral regurgitation. The usefulness of previously described corrective techniques depends on the specific origin of the anomalous artery.<sup>2</sup> We present the case of a 19-year-old woman who was diagnosed with a left coronary artery (LCA) that originated from the anterior-facing sinus of the pulmonary artery (PA). We describe how we modified a previously reported surgical technique and thereby overcame the long distance between the patient's aorta and the anomalous artery.

**Key words:** Anastomosis, surgical/methods; cardiac surgical procedures/methods; coronary vessel anomalies/surgery; pulmonary artery/abnormalities/surgery

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## Case Summary

In November 2009, a 19-year-old woman had a ventricular fibrillation cardiac arrest after moderate exertion. She underwent cardioversion and resuscitation by paramedics and had no obvious neurologic sequelae. Upon her admission to our hospital, the diagnosis was acute anterior myocardial infarction. The patient's cardiac troponin level was only slightly elevated. An echocardiogram showed a normal mitral valve and only mildly impaired ventricular function, due to anteroapical hypokinesia. A coronary angiogram confirmed our suspicion of ALCAPA. Magnetic resonance images revealed a dilated LCA originating from the anterolateral aspect of the pulmonary trunk, together with several collateral vessels between the right and left coronary arteries (Fig. 1). The patient was diagnosed with Bland-White-Garland syndrome and was scheduled for surgery.

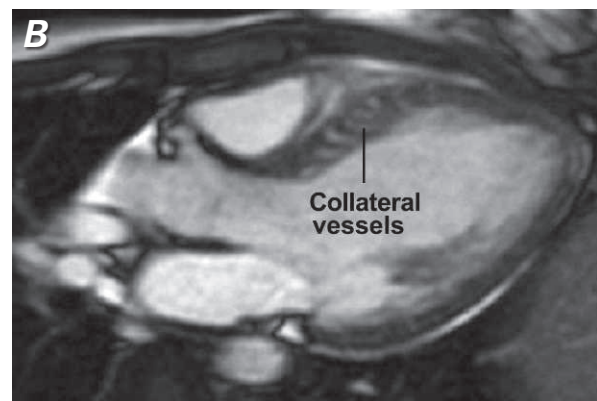
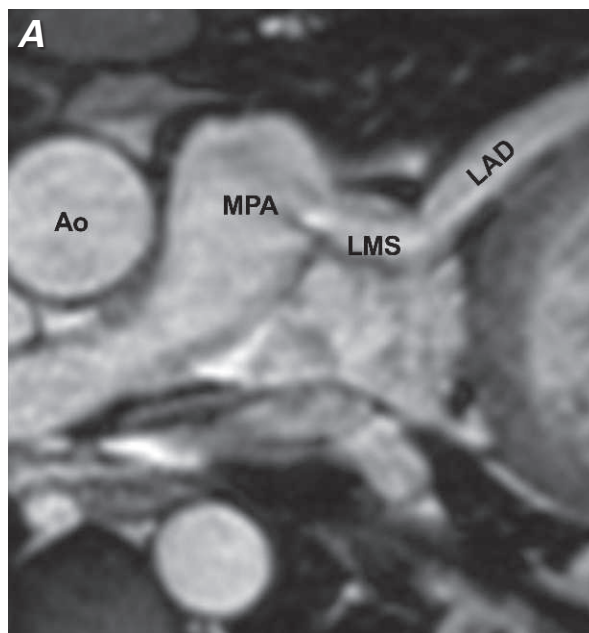
## Surgical Technique

We instituted mildly hypothermic cardiopulmonary bypass through a median sternotomy. Myocardial protection was achieved with use of cold-blood cardioplegic solution, first administered via the aortic root and main PA after the snaring of both PA branches and thereafter by direct injection. The LCA was dilated to approximately 6.3 mm and originated from the anterolateral aspect of the main PA. The tortuous right coronary artery was dilated to 8 mm (Fig. 2A). Both ventricles were moderately dilated.

The origin of the anomalous artery was resected, along with a long strip of PA extending medially. Similarly, a transverse strip of aorta was excised, and the hinge of the flap was placed anteriorly toward the PA (Fig. 2B). The PA was patched with a bovine pericardial strip and continuous 4-0 Prolene sutures, from the anterolateral sinus to the medial aspect of the PA (Fig. 2C). The strip of aortic wall was brought across so that its distal edge could be anastomosed to the posterior edge of the button of the LCA, and an anastomosis was constructed with use of 5-0 Prolene suture (Fig. 2C). A long sleeve was thus created with the PA strip anterior and the aortic flap posterior. We continued suturing until the new coronary artery was connected to the ascending aorta. We then patched the aorta with an additional strip of bovine pericardium. As a result, the

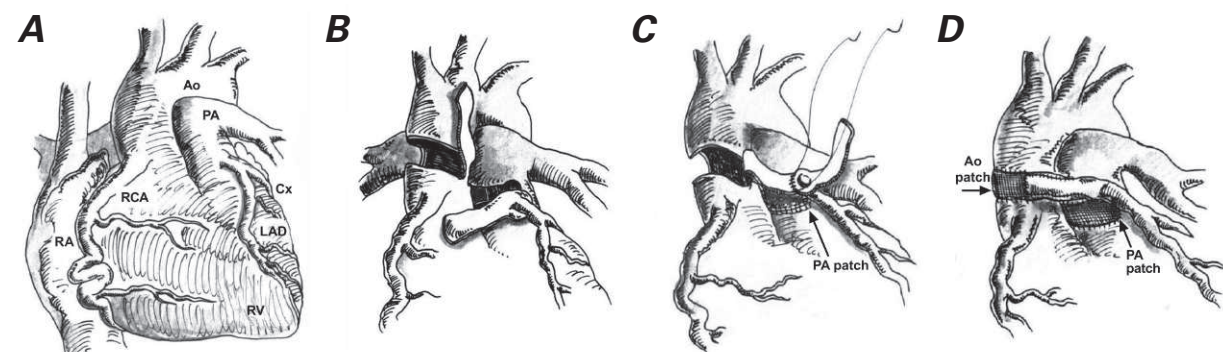
newly constructed left main system originated from the anterior aspect of the ascending aorta, 5 mm above the sinotubular junction (Fig. 2D). A postoperative echocardiogram showed preserved LV function.

The patient recovered rapidly and was discharged from the hospital on the 7th postoperative day. Upon follow-up examination 6 weeks later, she was asymptomatic. Two years after the surgery, a cardiac magnetic resonance angiogram showed a patent left coronary anastomosis without kinking, the patient's LV function was good (Fig. 3), and she was asymptomatic. The diameter of the right coronary artery was 6.8 mm. The proximal LCA appeared to be slightly more dilated than preoperatively, by approximately 0.5 mm; however, this might not have been a true dilation. Three years postoperatively, the patient remained asymptomatic.



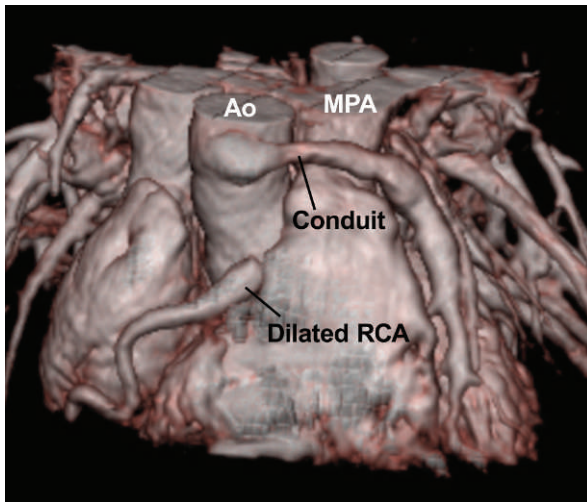
**Fig. 1** Cardiac magnetic resonance images. **A)** Axial oblique curved multiplanar reformatted image from a 3-dimensional whole-heart navigator sequence shows the left main stem (LMS) arising from the main pulmonary artery (MPA). The left anterior descending coronary artery (LAD) is substantially dilated. **B)** Collateral vessels between the right and left coronary arteries are dilated.

Ao = aorta



**Fig. 2** **A)** The dilated left anterior descending coronary artery (LAD) originates from the anterolateral sinus of the pulmonary artery (PA). The right coronary artery (RCA) is dilated. **B)** The left main button is resected along with a strip of PA, and a strip of aortic wall is resected above the sinotubular junction. **C)** The strips of PA and aorta are anastomosed to create a sleeve. The PA is repaired with bovine pericardium. **D)** Bovine pericardium is used to patch the aorta.

Ao = aorta; Cx = left circumflex coronary artery; RA = right atrium; RV = right ventricle



**Fig. 3** Postoperative volume-rendered magnetic resonance image from a 3-dimensional whole-heart sequence shows the newly constructed surgical conduit with no narrowing. The right coronary artery (RCA) remains dilated.

Ao = aorta; MPA = main pulmonary artery

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## Discussion

Surgical intervention—the definitive treatment for Bland-White-Garland syndrome—is challenging. The risk of postoperative heart failure and death depends on the patient's age and mode of presentation.<sup>3</sup> Different repair techniques have been described. One method is simple ligation of the anomalous artery to avoid steal and enable perfusion of the LV by collateral vessels; another method is to create a dual coronary system.<sup>3</sup> Currently, it is widely accepted that the best outcomes result from creating a dual coronary system by directly implanting the anomalous artery onto the ascending aorta.<sup>4</sup> However, when the LCA originates from the left side of the PA or follows an intramural course, direct reimplantation can be difficult.

In our patient, the origin of the anomalous artery was not juxtacommissural, nor was its course intramural; however, anatomic restrictions precluded direct reimplantation or the creation of an intrapulmonary tunnel. Our technique of creating a sleeve with a flap of PA strip and aorta in an anterior position is a modification of the technique described by Sodian and colleagues.<sup>5</sup> Creating a sleeve with autologous tissue advantageously bridged the anatomic gap without placing any tension on the anastomosis and enabled tissue growth. Our patient was asymptomatic after 3 years, and the new vessel remained patent. We think that our modified technique served as a valuable surgical alternative to correct the defect and maintain long-term patency, and that it might prove useful to others in similar circumstances.