

Bland - White - Garland Syndrome confirmed by dual source computed tomography angiography

V Mendoza-Rodríguez, L Llerena, E Olivares-Aquiles, M Sollet, J Valiente – Mustelier

Citation

V Mendoza-Rodríguez, L Llerena, E Olivares-Aquiles, M Sollet, J Valiente – Mustelier. *Bland - White - Garland Syndrome confirmed by dual source computed tomography angiography*. The Internet Journal of Cardiology. 2008 Volume 7 Number 1.

Abstract

A 16 years old woman, with typical chest pain and electrocardiogram changes was sent to our Institute to rule out anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) suspected by transthoracic echocardiography. Dual source computed tomography was performed and the diagnosis of Bland - White - Garland Syndrome with marked intercoronary collateral arteries and retrograde filling of the left coronary arterial system was done.

INTRODUCTION

In 1885, Brooks was the first to show that coronary arteries may anomalously originate from the PA (1). ALCAPA, otherwise known as Bland-White-Garland syndrome, is a rare congenital defect that accounts for 0.25–0.5% of all congenital heart defects (2). The diagnosis is suspected by clinical history,

electrocardiographic features, ischemia revealed in exercise treadmill testing or stress myocardial perfusion imaging, but the diagnosis is really established by angiographic study. Currently, color Doppler flow mapping might help to simplify the diagnosis in some cases. Although conventional coronary angiography is the gold standard for the diagnosis of coronary stenosis, the diagnosis accuracy of coronary anomalies is near 100% by Magnetic Resonance or multidetector computer tomography (3). We report a 16 years old woman with ALCAPA diagnosed by dual source computed tomography angiography.

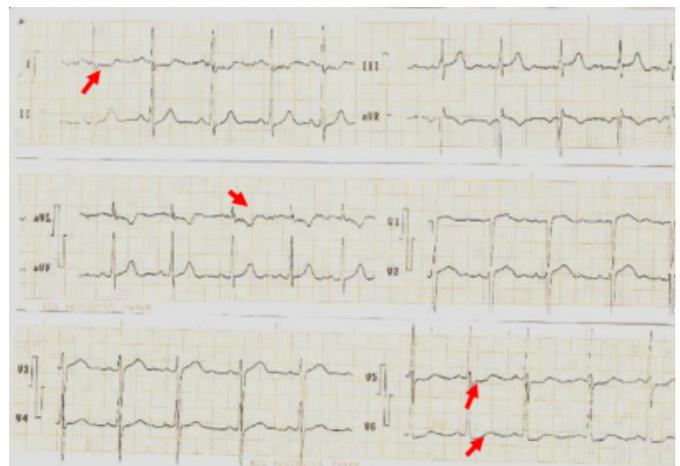
CASE REPORT

A 16 years old woman with history of typical chest pain and shortness of breath in exercise was first admitted in other Hospital and sent to our Institute in order to perform dual source computed tomography angiography. Her physical examination and chest X-Ray were normal. The electrocardiogram at rest revealed ST depression in D1, V5 and V6 leads and T wave inversion in aVL lead (Figure 1). An exercise treadmill test (Bruce protocol) revealed 2-mm

ST depression in anterolateral wall of the heart.

Figure 1

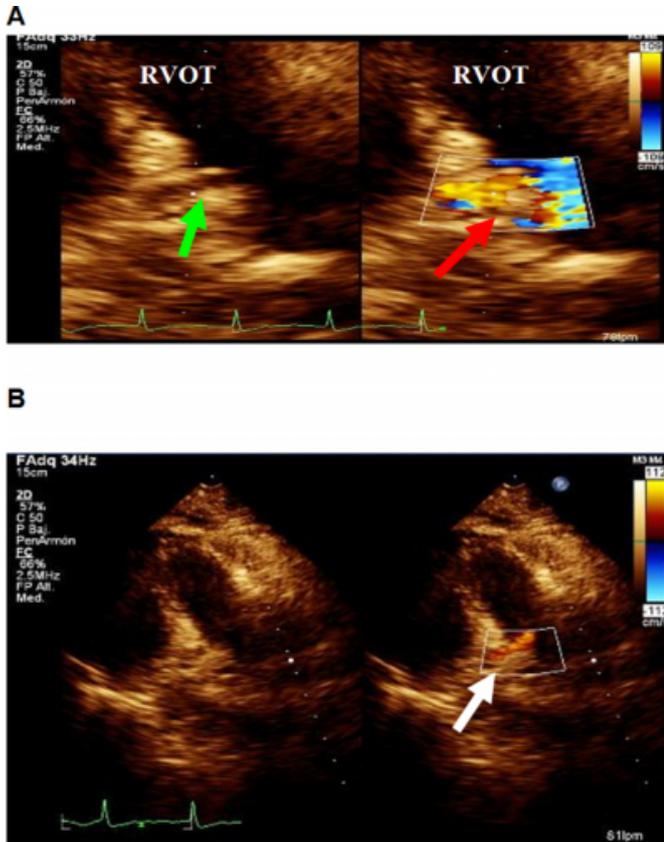
Figure 1. Electrocardiogram. ST segment depression in D1, V5 and V6 and T wave inversion in aVL (red arrows)



The transthoracic 2D echocardiography showed a mild reduced contractile apical wall, normal systolic left ventricular function, no abnormal echogenicity of the papillary muscles walls. A vessel with the characteristics of the left coronary artery emerging from the pulmonary artery trunk was seen (Figure 2A).

Figure 2

Figure 2. Echocardiography. A. In the right ventricular outflow tract (RVOT) view, a vessel (green arrow) is emerging from the posteromedial aspect of the pulmonary artery trunk just near to the pulmonary valve. B. A reliable flow signals were obtained with Doppler (white arrow)



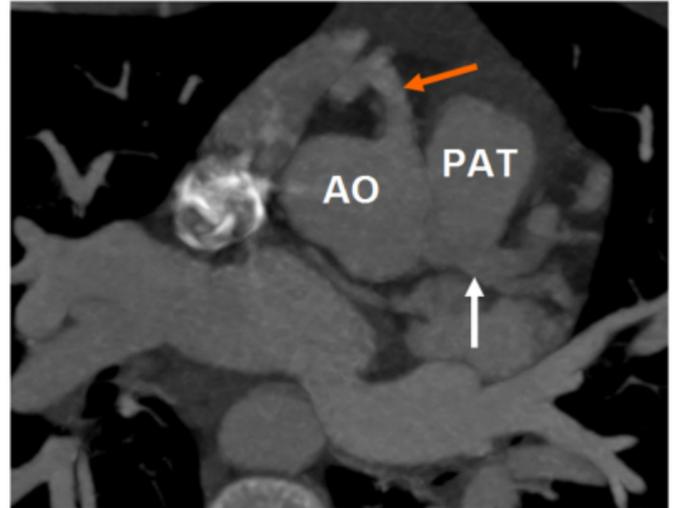
and abnormal flow from the left coronary artery to the pulmonary trunk was visible by the color Doppler mapping (Figure 2A red arrow and figure 2B white arrow).

The patient was sent to our Tomographic Department and studied in the dual source computed tomography, Somatom Definition, Siemens, Forchheim Germany, to confirm the diagnosis.

An enhanced contrast chest multidetector computed tomography was performed and confirmed the diagnose of left coronary artery origin from the pulmonary trunk (Figure 3).

Figure 3

Figure 3. Dual source computed tomography angiography. Axial view. Left coronary artery (white arrow) is emerging from pulmonary artery trunk (PAT) and right coronary artery (red arrow) emerging from aorta (AO).



An enormously dilated, tortuous and dominant right coronary artery (Figure 4) emerge from the right aortic sinus, with a large number of collateral vessels (Figure 5) feeding the left coronary system, which showed enlarged and tortuous vessels.

Figure 4

Figure 4. DSCT angiography. Inspace image. Left coronary artery (green arrow) is emerging from pulmonary artery trunk (PAT) and right coronary artery (white arrow) emerging from right coronary sinus of the aorta

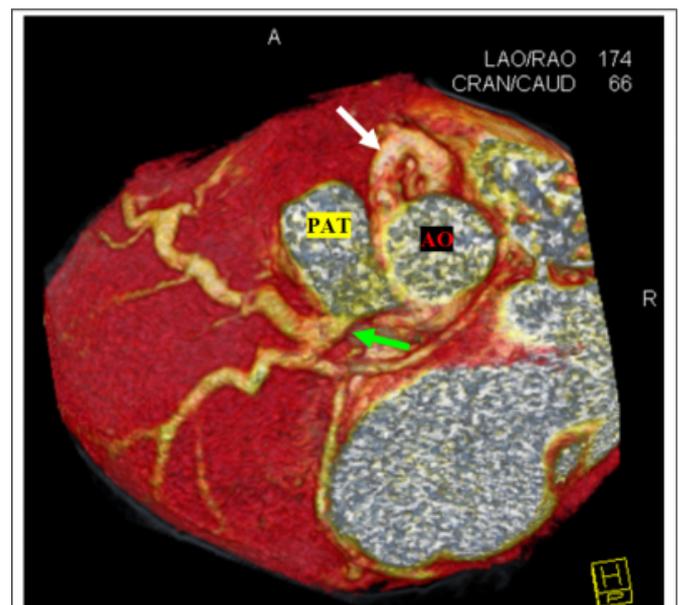


Figure 5

Figure 5. DSCT angiography. Inspace image. Intercoronary collateral circulation (arrows). A tortuous right coronary artery (red arrow) feeding the left coronary artery.



The patient was sent to the cardiovascular surgeon for treatment and Takeuchi surgery was performed. The evolution has been satisfactory.

DISCUSSION

Origin of the left coronary artery from the pulmonary artery, or Bland-White-Garland syndrome, is a rare anomaly that accounts for 0.5% of congenital malformations (4). It is frequently lethal in children and adults (5).

It most often presents as an isolated defect, but in 5% of cases it may be associated with other cardiac anomalies, including atrial septal defect, ventricular septal defect, and aortic coarctation (6).

Symptoms usually occur in infants after they are 1–2 months old because of left-to-right shunting from the higher pressure left coronary arterial system to the lower pressure pulmonary arterial system. This most often results in death due to circulatory insufficiency from left ventricular dysfunction or mitral valve incompetence, myocardial infarction, or life-threatening cardiac dysrhythmias. Before they are 1 month old, however, physiologic pulmonary arterial hypertension tends to preserve anterograde blood flow within the left coronary artery (LCA), accounting for the usual lack of symptomatology in this age group. (7) Without treatment, approximately 90% of infants die within the first year of life

(8). Rarely, however, they survive into adulthood with clinical presentations varying from symptomatic chronic mitral insufficiency or global ischemic cardiomyopathy to little or no symptomatology. Furthermore, the risk for sudden cardiac death due to ischemic malignant ventricular dysrhythmias exists even in asymptomatic adult patients (8, 9). Factors that may lead to survival beyond infancy include the development of abundant intercoronary collateral arteries, an alteration in hemodynamics that encourages anterograde blood flow into the left coronary arterial tree, and a reduction in the area of the left ventricular myocardium supplied by the LCA (8).

In our patient we saw a big intercoronary collateral circulation between the right coronary and the left coronary to supply the left ventricle flow.

The presence of collateral circulation between coronary arteries and systemic extracardiac vessels (9), mainly the bronchial arteries, is sporadically described in ALCAPA (10, 11). Along with the more commonly described intercoronary collaterals, these more rarely described systemic collaterals certainly contribute to survival with ALCAPA into adulthood; therefore, concern about the role of these systemic collaterals may seem meaningless from a practical point of view.

Conventional angiography can detect and depict the course of anomalous coronary arteries, although this sometimes can be difficult to perform or interpret. This procedure is invasive and severe complications may occur.

Most clinicians recognize the need for surgical treatment, due to reports of recurrent arrhythmias, cardiac failure, or sudden death (5, 12). Various surgical approaches have been described (1, 5, 7, 13). Frequently, the conventional angiographic study (right coronary arteriography or angiography in the pulmonary trunk) does not show the correct anatomy of the left coronary artery, due to opacification of large “collateral-like” vessels. For this reason, the surgical technique is frequently decided during the surgery, increasing mortality and morbidity.

Magnetic resonance and multislice computed tomography angiography have a very high accuracy to detect these anomalies and the cardiac anatomy (3). The diagnosis with these techniques may help the surgeons and the prognosis will be better for the patients.

References

1. Purut CM, Sabiston DC. Origin of the coronary artery

- from the pulmonary artery in older adults. *J Thorac Cardiovasc Surg* 1991; 102: 566-70.
2. Pfannschmidt J, Ruskowski H, de Vivie ER. Bland White Garland syndrome Clinical aspects, diagnosis, therapy. *Klin Padiatr* 1992; 204:328-334
 3. Schroeder S, Achenbach S, Bengel F, Burgstahler C, Cademartiri F, de Feyter P, George R, Kaufmann P, Kopp AF, Knuuti J, Ropers D, Schuijf J, Tops L F and Bax JJ. Cardiac computed tomography: indications, applications, limitations, and training requirements. *Eur Heart J* 2008; 29:531-556
 4. Saeed BT, Rosin MD, Murray RG. Successful operation in an old survivor of anomalous origin of the left coronary artery from the pulmonary trunk (Bland- White-Garland Syndrome). *Br Heart J* 1994; 71: 193-5.
 5. Moodie DS, Fyfe D, Gill CC, et al. Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland Syndrome) in adult patients: Long-term follow-up after surgery. *Am Heart J* 1983; 106: 381-8.
 6. Takimura CK, Nakamoto A, Hotta VT, Campos MF, Malamo M, Otsubo R. Anomalous origin of the left coronary artery from the pulmonary artery: report of an adult case. *Arq Bras Cardiol* 2002; 78: 309-314.
 7. Karolczak MA, Wieteska J, Bec L, Madry W. Anomalous origin of the left coronary artery (LCA) from pulmonary trunk (Bland-White-Garland syndrome) with systemic collateral supply to LCA. *Med Sci Monit* 2001; 7:755 -758.
 8. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. *Circulation* 1968; 38: 403-425.
 9. Moberg A. Anastomoses between extracardiac vessels and coronary arteries. I. Via bronchial arteries. Post-mortem angiographic study in adults and newborn infants. *Acta Radiol Diagn (Stockh)* 1967; 6:177-192.
 10. Ishihata T, Takeda H, Katohno E, et al. An adult case of Bland-White-Garland syndrome with collaterals from the bronchial artery. *Heart Vessels* 1994; 9:218-222.
 11. Karolczak MA, Wieteska J, Bec L, Madry W. Anomalous origin of the left coronary artery (LCA) from pulmonary artery (Bland-White-Garland syndrome) with systemic collateral supply to LCA. *Med Sci Monit* 2001; 7:755-758.
 12. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology. Based on a review of 140 cases with seven further cases. *Circulation* 1968; 38: 403-25.
 13. Oliveira SA, Diament J, Carvalho VB, Arie S, Macruz R, Zerbini EJ. Anomalous origin of the left coronary artery from the pulmonary artery. Surgical repair of on unused form. *J Cardiovasc Surg* 1977; 18: 599-605.

Author Information

V Mendoza-Rodríguez, MD

Institute of Cardiology and Cardiovascular Surgery, Habana, Cuba

LR Llerena, MD, PhD

Institute of Cardiology and Cardiovascular Surgery, Habana, Cuba

EW Olivares-Aquiles, MD

Institute of Cardiology and Cardiovascular Surgery, Habana, Cuba

M Sollet, MD

William Soler Hospital. Pediatric Cardiology Department

J Valiente – Mustelier, MD

Institute of Cardiology and Cardiovascular Surgery, Habana, Cuba