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Brief Report

Anomalous left coronary artery arising from the right pulmonary artery in association with coarctation of the aorta

Xuecun Liang, Lin Wu

Cardiovascular Center, Children's Hospital of Fudan University, Shanghai, PR China

Abstract In this study, we describe a very rare case of anomalous left coronary artery arising from the right pulmonary artery in association with coarctation of the aorta. A 3-month-old boy presented with refractory congestive heart failure since 20 days after birth. The initial echocardiography suggested the diagnosis of left coronary artery-to-right pulmonary artery fistula associated with coarctation; however, selective coronary angiography demonstrated the rare anomaly of the left coronary artery arising from the right pulmonary artery. Subsequently, he underwent successful transcatheter balloon angioplasty for aortic coarctation and surgical repair of left coronary artery re-implantation.

Keywords: Coronary anomaly; pulmonary artery; echocardiography; angiography

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ANOMALOUS LEFT CORONARY ARTERY ARISING FROM the pulmonary artery is a rather rare congenital anomaly. It occurs in ~1/300,000 live births and represents <0.5% of CHDs. It was first described anatomically in 1885 by Brook[1] and clinically in 1933 by Bland et al.[2] Therefore, it is also known as Bland–White–Garland syndrome. Typically, the left coronary artery arises directly from the main pulmonary trunk adjacent to the normal aortic origin as an isolated malformation. In this study, we report an extremely rare case of anomalous left coronary artery arising from the proximal right pulmonary artery in association with coarctation of the aorta.

A 3-month-old boy, who presented with dyspnoea since 20 days after birth, was transferred to our hospital for refractory and progressive congestive heart failure by ambulance. Electrocardiography showed right ventricular hypertrophy with myocardial strain (Fig 1a). Echocardiography revealed the presence of coarctation and a significantly enlarged left ventricle with severely depressed systolic function. In the modified parasternal short-axis view, the left coronary artery seemed to be arising from the aortic root with continuous flow to the right pulmonary artery (Fig 1b and c). Therefore, the suggestive diagnosis of left coronary artery-to-right pulmonary artery fistula was made; however, at cardiac catheterisation, aortic root angiography showed the dilated right coronary artery arising from the right aortic sinus without the normal origin of left coronary artery being seen. Selective right coronary artery angiography demonstrated that the left coronary artery was supplied by the right coronary artery through major collaterals between the right coronary artery and the left anterior descending artery and a communication between the right coronary artery and circumflex arteries, finally with retrograde filling of the proximal right pulmonary artery (Fig 2, Supplementary video 1). Transcatheter angioplasty was performed using a 6-mm balloon (Boston Scientific, Natick, MA, United States of America) for the discrete coarctation. After angioplasty, the systolic pressure gradient across the coarctation decreased from 30 to 6 mmHg, and the diameter of the coarctation site increased from 3.24 to 4.53 mm. Surgical repair of the left coronary artery re-implantation in order to establish two-coronary-artery circulation was performed 6 days after catheterisation.

Correspondence to: L. Wu, Cardiovascular Center, Children's Hospital of Fudan University, 399 Wan Yuan Road, Shanghai 201102, PR China. Tel: +86 216 493 2800; Fax: +86 216 493 2802; E-mail: wulin0609@foxmail.com

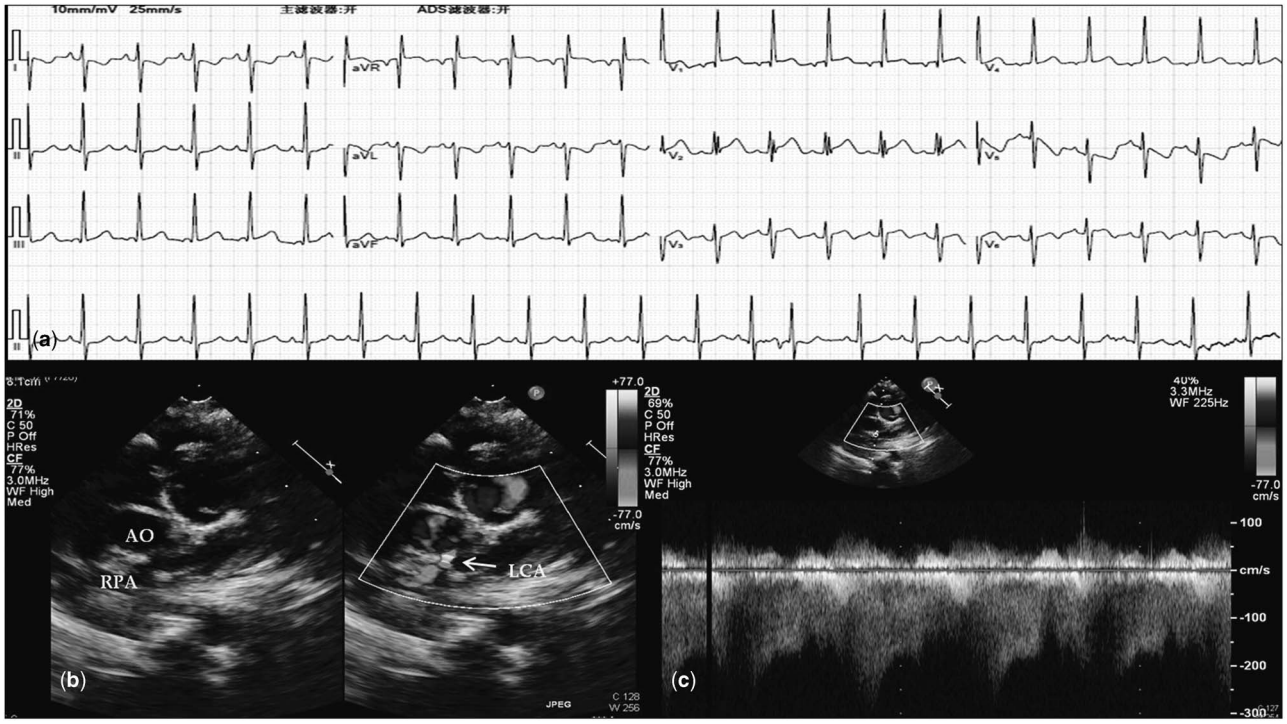

Figure 1.
*A 12-lead electrocardiogram showing right ventricular hypertrophy with myocardial strain. No evidence of myocardial ischaemia or infarct is demonstrated (**a**). On two-dimensional echocardiography, the high left oblique parasternal short-axis view of the RPA revealed that a coronary flow drained into the RPA during diastole (**b**). A pulse-wave Doppler spectrum demonstrating the continuous coronary artery flow pattern (**c**). AO = aortic artery; LCA = left coronary artery; RPA = right pulmonary artery.*

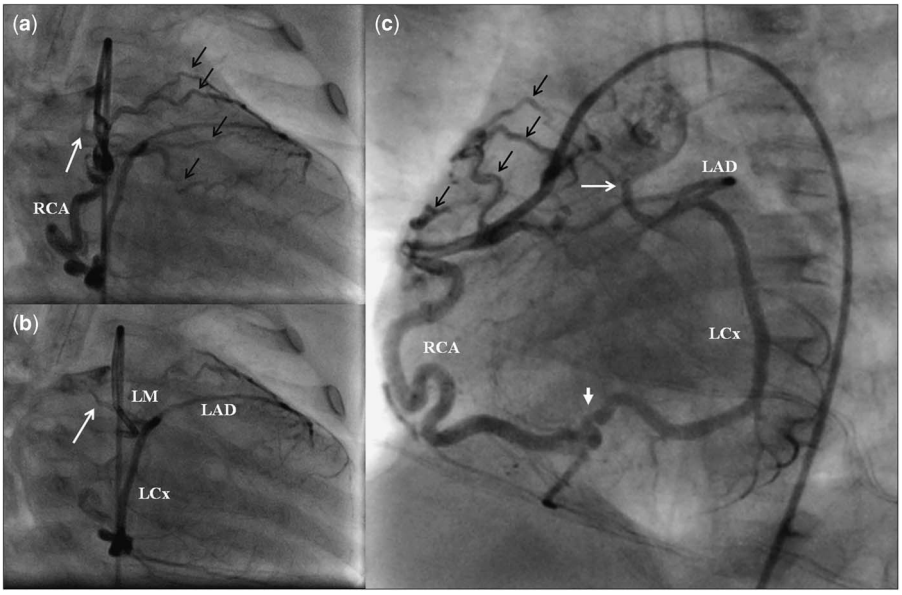

Figure 2.
Selective right coronary artery angiogram showing mildly dilated and tortuous RCA supplying the LCA through major collaterals (black arrow) between the RCA and the LAD, and communication between the RCA and the LCx (short arrow). The LM retrograde flows into the proximal right pulmonary artery (white arrow). LAD = left anterior descending coronary artery; LCA = left coronary artery; LCx = left circumflex branch; LM = left main coronary artery; RCA = right coronary artery.

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The procedure was eventful and echocardiography showed improved left ventricular systolic function at the 2-week follow-up after surgical repair.

In retrospection of this case, two points need to be addressed. First, the severity of myocardial ischaemia is not proportional to that of left ventricular dysfunction. Usually, abnormal Q-waves exist in leads I, avL, and V_4 through V_6, demonstrating the anterolateral myocardial infarct in the patient with anomalous left coronary artery arising from the pulmonary artery. In this case, there was no evidence of myocardial ischaemia on the electrocardiography strip, whereas the left ventricular failure was significant. The reason for the discrepancy is that the presence of coronary collaterals alleviates the degree of myocardial ischaemia, and the impaired left ventricular function attributes not only to anomalous left coronary artery arising from the pulmonary artery but also to coarctation. Second, the fistula suggested by echocardiography actually is the orifice of the left coronary artery arising from the right pulmonary artery, with retrograde continuous flow from the left coronary artery to the right pulmonary artery. With regard to young infants with significant left ventricular dilatation and dysfunction, anomalous left coronary artery arising from the pulmonary artery is always one of the important differential diagnoses during echocardiography investigation. In this case, because the origin of the left coronary artery was not seen at the main pulmonary trunk, anomalous left coronary artery arising from the pulmonary artery was ruled out in our initial evaluation of the echocardiography. Therefore, this case reminds us to keep it in mind that the left coronary artery could originate from the right pulmonary artery as a special rare type of anomalous left coronary artery arising from the pulmonary artery.

In conclusion, clinicians should be aware that the left coronary artery can rarely arise from the right pulmonary artery instead of the pulmonary trunk in patients with anomalous left coronary artery arising from the pulmonary artery.[3,4] Thorough echocardiography evaluation is crucial for the early diagnosis and favourable recovery of left ventricular function after surgical repair.

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Conflicts of Interest

None.

Ethical Standards

This study was approved by the Ethics Committee of Children's Hospital of Fudan University.

Supplementary material

For supplementary material referred to in this article, please visit http://dx.doi.org/10.1017/S1047951111500195X

References

1. Brooks HS. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effect of this anomaly in producing cirsoid dilatation of the vessels. J Anat Physiol 1886; 20: 26–29.
2. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. Am Heart J 1933; 8: 787–801.
3. Bhatia AK, Kreeger J, Slesnick T, Kogon B, Kim DW. Infant with anomalous left coronary artery arising from the right pulmonary artery and ventricular septal defect. Circulation 2012; 127: e544–e546.
4. Zheng JY, Ding WH, Xiao YY, Jin M. Anomalous origin of the left coronary artery from the pulmonary artery in children: 15 years experience. Pediatr Cardiol 2011; 32: 24–31.