ESC European Society of Cardiology

European Heart Journal - Case Reports (2020) **4**, 1–4
doi:10.1093/ehjcr/ytaa293

CASE REPORT
Congenital heart disease

One patient with an anomalous origin of the left pulmonary artery directly from the right ventricle: a case report

Xiaomin He[1], Zhifang Zhang[2], Jinghao Zheng[1]*, and Zhongqun Zhu[1]*

[1]Department of Cardiothoracic Surgery, Shanghai Children's Medical Center, Affiliated to Shanghai Jiao Tong University School of Medicine, 1678 Dongfang Road, Shanghai 200127, China; [2]Department of Echocardiography, Shanghai Children's Medical Center, Affiliated to Shanghai Jiao Tong University School of Medicine, 1678 Dongfang Road, Shanghai 200127, China; and

Received 10 March 2020; first decision 26 March 2020; accepted 4 August 2020; online publish-ahead-of-print 5 November 2020

Background The anomalous origin of one pulmonary artery is a rare malformation, which so far has mainly been found as an anomalous origin from a different site of the aorta, accounting for 0.12% of all congenital heart diseases. This case report introduced a very rare case of the anomalous origin of one pulmonary artery which had never reported in the clinic.

Case summary A 2-year-old boy with a 6-month history of shortness of breath and recurrent respiratory infection, was diagnosed left pulmonary artery (LPA) directly arising from the right ventricle by transthoracic echocardiography and multidetector computed tomography without a deletion in the region of 22q11. Eventually, the LPA was further conformed that arised from the right ventricle during the operation, and was corrected with a well clinical outcome.

Discussion The surgical technique for repair of this anomalous LPA was not difficult in our case. However, the embryonic development of the present case still could not be completely explained by the current embryologic postulates since it was a new malformation that never reported. Due to its rarity, there is still much to learn about the origin and development of the pulmonary arteries that possibly develop prenatally.

Keywords A congenital heart surgeon • Congenital heart disease • Great vessels anomalies • Pulmonary arteries • Case report

Learning points

- The anomalous origin of one pulmonary artery is a rare malformation
- The left pulmonary artery directly arising from the right ventricle is very rare, and such a finding has never reported in the clinic
- Accurate diagnosis and appropriate surgical treatment has well clinical outcome for that anomalous origin of one pulmonary artery
- Due to its rarity, there is still much to learn about the origin and development of the pulmonary arteries that possibly develop prenatally

* Corresponding authors. Tel: +86 21 38626161, Fax: +86 21 58393915, Email: zhengjh210@163.com (J.Z.); Tel: +86 21 38626161, Fax: +86 21 58393915, Email: zhuzhongqun@scmc.com.cn (Z.Z.)
Handling Editor: Inga Voges
Peer-reviewers: Filippo Puricelli; Golnaz Houshmand
Compliance Editor: Amir Aziz

Introduction

To the best of our knowledge, an anomalous origin of one pulmonary artery, which could originate from the right pulmonary artery (RPA), ascending aorta (AAO), innominate artery, or descending aorta, is a very rare congenital cardiac malformation accounting for 0.12% of all congenital heart diseases.[1–3] However, we here report a case of left pulmonary artery (LPA) directly arising from the right ventricle in a 2-year-old boy, and such a finding has never been reported in the clinic.

Timeline

Day 1	Patient presented in the outpatient department with a 6-month history of shortness of breath and recurrent respiratory infection. Transthoracic echocardiography (TTE) found that the origin of the left pulmonary artery (LPA) was abnormal
Day 3	Multidetector computed tomography (MDCT) further demonstrated the anomalous origin of the LPA arising from the right ventricle with stenosis, and a large lateral branch arising from the descending aorta to supply the left lung blood
Day 5	The genetic testing did not identify a deletion in the region of 22q11
Day 6	The surgical correction was performed, and the LPA was further conformed arising from the right ventricle during the operation
Day 13	The patient was successfully discharged on postoperative Day 7
Day 37	The whole corrected course and morphometry of pulmonary artery were confirmed by TTE and MDCT 1 month after operation
Month 23	The patient has been followed up without any symptoms

Case presentation

A 2-year-old boy (body surface area 1.8 m^2) came to our hospital with a 6-month history of shortness of breath and recurrent respiratory infection. Initial examination showed no obvious positive clinical signs. Transthoracic echocardiography (TTE) found that the origin of the LPA was abnormal. There was a blood vessel arising from the right side of the pulmonary annulus, which went across the main pulmonary artery (MPA) to the left pulmonary hilum with an inner diameter of 3.5 mm (*Figure 1A*), while the RPA was normal with approximately a 8 mm diameter. The *z*-value of the LPA and RPA was -3.16 and 2.4, respectively. Although biphasic blood flow in this vessel could be observed by TTE, the proximal orificium of this vessel was narrow so that the amount of regurgitation to the right ventricle was very limited (*Figure 1B*). Other findings included left atrium and left ventricle enlargement, right aortic arch (*Figure 1C*). Multidetector computed tomography (MDCT) further demonstrated the anomalous origin of the LPA arising from the right ventricle with stenosis, and a large lateral branch arising from the descending aorta to supply the left lung blood (*Figure 1D–F*). The patient had no other intracardiac or extracardiac malformations. Besides, there was no relevant family history, and subsequent genetic testing did not identify a deletion in the region of 22q11.

We performed a surgical correction for this patient. First, the large lateral branch was found and ligated through the left sixth intercostal space thoracotomy without any effect on oxygenation. Then, a midline sternotomy was performed, and operative inspection finally demonstrated the above diagnosis. The LPA was seen arising from the root of MPA near the AAO and crossing the anterior wall of the MPA to the upper left, finally entering into the left pulmonary hilum with approximately a 3.5 mm diameter (*Figure 2A*). During the cardiopulmonary bypass, we found that LPA and MPA had a common wall without connection (*Figure 2B*). The LPA was actually arising from the right ventricle without valvular tissue (*Figure 2C*). The proximal orificium of the LPA was closed with interrupted sutures by the autologous pericardium (*Figure 2D*). The connection of the LPA and MPA was completed through the incision of the common wall (*Figure 2E*). Then, the anterior wall of the LPA was enlarged to its bifurcation with an autologous pericardium patch (*Figure 2F*). The patient successfully received anatomic correction and discharged in postoperative Day 7. The whole corrected course and morphometry of pulmonary artery were confirmed by TTE and MDCT 1 month after operation (*Figure 2G–I*). So far, the patient has been followed up without any symptoms for 23 months.

Discussion

The arterial trunks are formed within the distal part of the outflow tract, subsequent to ongoing migration of cells into the arterial pole from the second heart field. Originally, the arterial trunks extend through the fourth pharyngeal arches, which will become the systemic vessels. Then the arterial trunks arise from the cranial part of the sac with the sixth arches, which give rise to the developing right and left pulmonary arteries.[4] The left sixth arch continues as the ductus arteriosus until after birth when the right sixth arch is absorbed completely. The anomalous origin of one pulmonary artery is a very rare malformation, which so far has mainly been found as an anomalous origin from different sites of the aorta.[5]

We found a case of LPA directly arising from the right ventricle, and to our knowledge, such a finding has never been reported in the clinic. There were several embryologic postulates for the anomalous origin of the LPA from the aorta (ALPA). ALPA might result from failure of the development of the left sixth arch and persistence of the left fifth arch.[3] However, these embryologic postulates did not explain the embryonic development of our case, where the LPA was directly connected to the right ventricle. Since the LPA was connected with the right ventricle, our case was also different from crossed pulmonary arteries, in which the LPA originated from MPA to the right and usually lied above the origin of the RPA.[6] Besides, our case could not be accurately described as a 'triarterial heart' since the

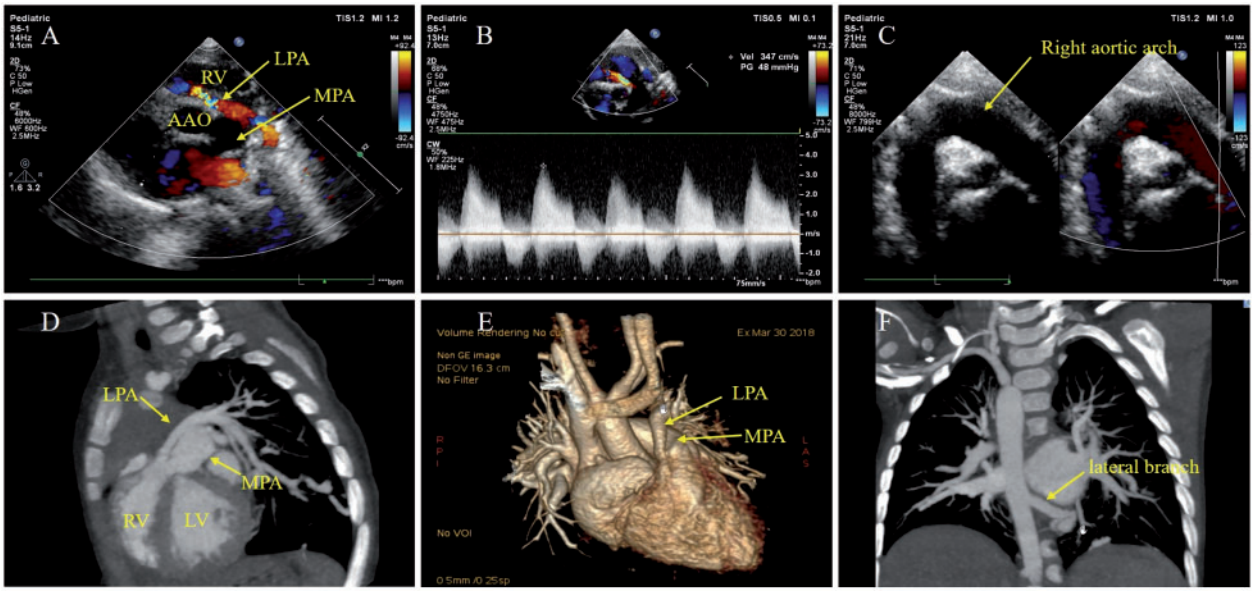

Figure 1 Anomalous origin of the left pulmonary artery from the right ventricle diagnosed by transthoracic echocardiography and multidetector computed tomography. (*A*) Transthoracic echocardiography showed that a blood vessel arising from the right side of main pulmonary artery and then across main pulmonary artery to the left pulmonary hilum. (*B*) Biphasic blood flow in this vessel was observed by transthoracic echocardiography. (*C*) Right aortic arch was observed by transthoracic echocardiography. (*D* and *E*) The left pulmonary artery arising from right ventricle with stenosis was demonstrated by multidetector computed tomography. (*F*) A large lateral branch arising from the descending aorta to supply the left lung blood by multidetector computed tomography. AAO, ascending aorta; LPA, left pulmonary artery; LV, left ventricle; MDCT, multidetector computed tomography; MPA, main pulmonary artery; RPA, right pulmonary artery; RV, right ventricle; TTE, transthoracic echocardiography.

LPA had no valve tissue and infundibulum of its own. Furthermore, this abnormal embryonic development might be different from conotruncal cardiac malformations, as the 22q11 chromosomal deletion did not exist in our case.

In addition, in our case we found that a large lateral branch arose from the descending aorta. It could be argued that the lateral branch supplying the blood flow to the left lung might be the anomalous origin of the LPA, since ALPA could also arise from the descending aorta as reported.[3,7] However, its branching, distribution, and morphology were more like an aortopulmonary collateral case, and the abnormal artery from the right ventricle is more likely to be the LPA. Besides, we found that the atrial and left ventricular dilation existed in this cardiac malformation, which may be related to the large lateral branch arising from the descending aorta, resulting in the increased blood flow back to the left atria.

Although the surgical technique for repair of this anomalous LPA was not difficult in our case, the embryonic development of the present case still could not be completely explained by the current embryologic postulates since it was a new malformation that never reported. Due to its rarity, there is still much to learn about the origin and development of the pulmonary arteries, the anomaly of which could possibly develop in prenatal stage.

Lead author biography

Xiaomin He is an Attending Physician, Medical Doctor in the Department of Cardiothoracic Surgery at Shanghai Children's Medical Center Affiliated to Shanghai Jiao Tong University School of Medicine. Dr He specializes in the surgical treatment and research in paediatric complex congenital heart disease, and also integration treatment including prenatal consultation, postnatal evaluation relating to congenital heart diseases.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

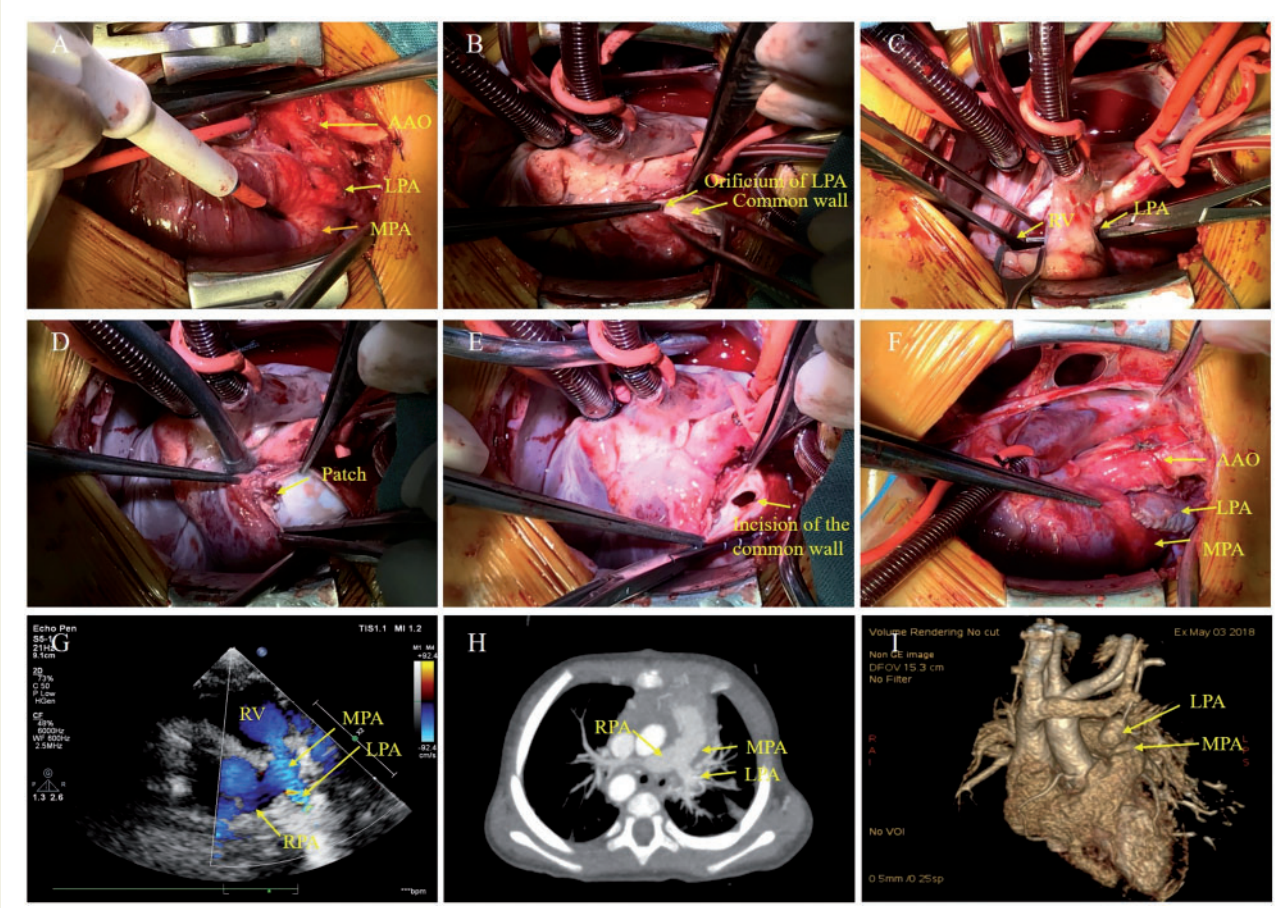

Figure 2 The left pulmonary artery arising from the right ventricle demonstrated during operation and followed up by transthoracic echocardiography and multidetector computed tomography postoperation. (*A*) Left pulmonary artery was seen arising from the root of the main pulmonary artery nearby the ascending aorta, and cross the anterior wall of main pulmonary artery to the upper left and came into the left pulmonary hilum during operation. (*B*) The arrow showed the proximal orificium of left pulmonary artery to the right ventricle and a common wall between the left pulmonary artery and main pulmonary artery. (*C*) The left pulmonary artery was actually connected with the right ventricle without valvular tissue. (*D*) The proximal orificium of left pulmonary artery was closed with interrupted sutures by autologous pericardium. (*E*) The connection of the left pulmonary artery and main pulmonary artery was completed through the incision of the common wall. (*F*) The anterior wall of left pulmonary artery was enlarged to its bifurcation with autologous pericardium patch. (*G*) Transthoracic echocardiography showed that left pulmonary artery was corrected and continued from main pulmonary artery. (*H* and *I*) Multidetector computed tomography demonstrated that left pulmonary artery was corrected and continued from main pulmonary artery postoperation. AAO, ascending aorta; LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery; RV, right ventricle.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

1. Yong MS, d'Udekem Y, Brizard CP, Robertson T, Robertson CF, Weintraub R et al. Surgical management of pulmonary artery sling in children. *J Thorac Cardiovasc Surg* 2013;**145**:1033–1039.
2. Cho S, Kim WH, Choi ES, Lee JR, Kim YJ. Surgical results of anomalous origin of one pulmonary artery branch from the ascending aorta. *Pediatr Cardiol* 2015;**36**: 1532–1538.
3. Gnanappa GK, Laohachai K, Orr Y, Ayer J. Isolated anomalous origin of left pulmonary artery from the descending aorta: an embryologic ambiguity. *Ann Thorac Surg* 2016;**102**:e439–e441.
4. Anderson RH, Mori S, Spicer DE, Brown NA, Mohun TJ. Development and morphology of the ventricular outflow tracts. *World J Pediatr Congenit Heart Surg* 2016;**7**:561–577.
5. Kutsche LM, Van Mierop LH. Anomalous origin of a pulmonary artery from the ascending aorta: associated anomalies and pathogenesis. *Am J Cardiol* 1988;**61**: 850–856.
6. Jue KL, Lockman IA, Edwards JE. Anomalous origins of pulmonary arteries from pulmonary trunk (crossed pulmonary arteries). *Am Heart J* 1966;**71**: 807–812.
7. Prifti E, Bonacchi M, Murzi B, Crucean A, Bernabei M, Luisi VS et al. Anomalous origin of the left pulmonary artery from the aorta. Our experience and literature review. *Heart Vessels* 2003;**18**:79–84.