Cardiology in the Young (2013), **23**, 466–469
doi:10.1017/S1047951112001308

Brief Report

Double outlet right ventricle with unilateral absence of left pulmonary artery

Murat Ugurlucan, Ahmet H. Arslan, Yahya Yildiz, Sibel Ay, Sertac Cicek

Anadolu Medical Center Hospital, Cardiovascular Surgery Clinic, Istanbul, Turkey

Abstract Congenital absence of unilateral pulmonary artery, either as a primary defect or in combination with other congenital cardiovascular malformations, is very rare. Double outlet right ventricle pathology in combination with unilateral absence of pulmonary artery has only been reported once in the literature. In this report, we present our experience with double outlet right ventricle with unilateral absence of left pulmonary artery in a 3-year-old female patient who underwent a palliative procedure and was scheduled for correction.

Keywords: Double outlet right ventricle; tetralogy of Fallot; ventricular septal defect; branch; single

Received: 28 March 2012; Accepted: 16 July 2012; First published online: 20 September 2012

Double outlet right ventricle, although not frequent among cardiac defects, is a well-known congenital cardiac malformation.[1,2] The pathology may be associated with pulmonary stenosis, atrial septal defect, anomalies of the aortic arch, patent ductus arteriosus, systemic or pulmonary venous return abnormalities, and atrioventricular canal defect.[1] Unilateral absence of a branch pulmonary artery, either the right or the left branch, is very rare. Double outlet right ventricle with unilateral absence of the left pulmonary artery has been reported once as a single case report in 1992.[3]

In this report, we present the case of a patient with double outlet right ventricle and absent left pulmonary artery together with our treatment strategy and post-operative course.

Case report

A 3-year-old female child living in a rural area was referred to our institution with the suspicion of a congenital heart disease due to cyanosis and growth retardation. She was 81 centimetres long and weighed 7.250 kilograms with a body mass index of 11.03 kilograms per metre square and 0.40 square metre body surface area. On admission, her room air oxygen saturation was 50%. There was pansystolic murmur at cardiac auscultation. Chest roentgenogram indicated increased cardiothoracic index. Echocardiography revealed double outlet right ventricle – origination of both arterial trunks from the right ventricle, more than 90% aortic override, and loss of aorto-mitral continuity – with pulmonary valvular and arterial stenosis. Pulmonary annulus sized 4.3 millimetres. The ventricular septal defect component was large, non-restrictive, and subaortic. The main and right pulmonary artery diameters were measured to be 5.5 and 5.3 millimetres, respectively; however, the left pulmonary artery could not be visualised. The Z-scores of pulmonary valve, main pulmonary artery, and the right pulmonary artery were calculated to be 8.59, 9.58, and 12.02, respectively. Further investigation of the pulmonary vasculature was performed with computerised tomography and cardiac catheterisation and both indicated the true absence of the left pulmonary artery (Figs 1 and 2). Moreover, pulmonary venous wedge angiography showed a pulmonary artery at the hilum of left lung (Fig 3).

We decided to perform a palliative procedure rather than correction. Through median sternotomy, the brachiocephalic trunk and right pulmonary

Correspondence to: Prof. Dr S. Cicek, MD, FACC, FACS, Anadolu Medical Center Hospital, Cardiovascular Surgery Clinic, Cumhuriyet Mahallesi, 2255 Sokak, No. 3, Gebze 41400, Kocaeli, Turkey. Tel: +90 262 678 50 67; Fax: +90 262 654 05 38; E-mail: sertac.cicek@anadolusaglik.org

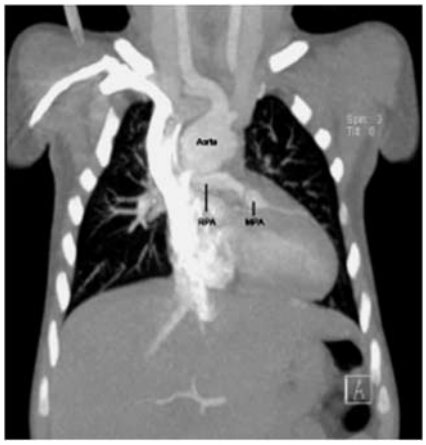

Figure 1.
Computerised tomography indicating the true absence of the left pulmonary artery. MPA = main pulmonary artery; RPA = right pulmonary artery.

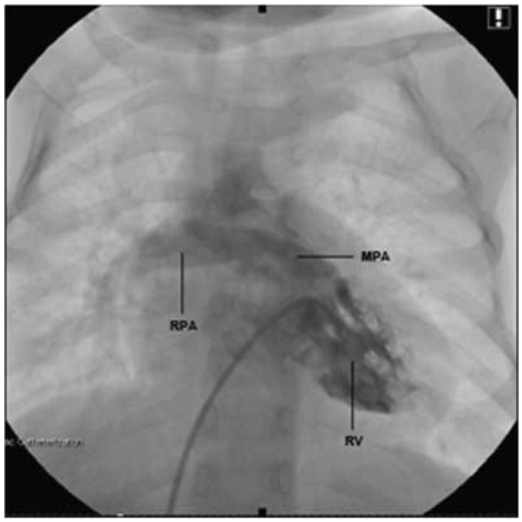

Figure 2.
Cardiac catheterisation showing right ventricular outflow tract, main and right pulmonary arteries, and absent left pulmonary artery. MPA = main pulmonary artery; RPA = right pulmonary artery; RV = right ventricle.

artery were exposed. A 4-millimetre expanded polytetrafluoroethylene graft (GoreTex, W.L. Gore & Associates Incorporation, Elkton, Maryland, United States of America) was anastomosed end to side to the brachiocephalic trunk. The ascending aorta and right atrium were cannulated and cardiopulmonary bypass was initiated. The other end of the graft was anastomosed end to side to the right pulmonary artery. Weaning off the cardiopulmonary bypass was uneventful. The shunt created 5-millimetre mercury systolic and 7-millimetre mercury diastolic gradients, increasing the oxygen saturation to 82% while still intubated and with

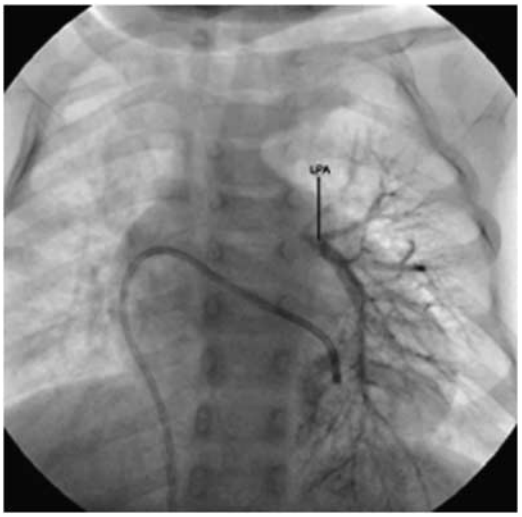

Figure 3.
Pulmonary venous wedge angiography showed a hilar left pulmonary artery. LPA = left pulmonary artery.

20% inspiratory oxygen. She was transferred to the intensive care unit and extubated after 8 hours. The post-operative course was complicated mainly because of overflow phenomenon and treated with inotropic support (5 micrograms per kilogram per minute of dopamine) for 3 days and forced diuresis with furosemide. Digoxin and aspirin were started.

Before discharge, she presented with a sudden drop of the oxygen saturation to 30% with dyspnoea on the 7th day. She was immediately intubated and a control echocardiography indicated shunt occlusion. She was taken to the operating theatre and the sternum was opened. The aorta and the right atrium were cannulated. Cooling was started, and during cooling a 9-millimetre-diameter pericardial roll was created. At 18°C, the heart was arrested with antegrade cold blood cardioplegia and circulatory arrest was initiated. The hilar pulmonary artery in the left lung was found. The pericardial roll was anastomosed end to side to the pulmonary artery at the hilum of the left lung with an 8.0 polypropylene suture using the single continuous anastomosis technique with extensive care in order to avoid damage to the fragile vasculature at this location. During the warming phase, the other end of the pericardial roll was anastomosed to the left side of the main pulmonary artery. The brachiocephalic trunk end of the polytetrafluoroethylene graft used during the previous operation was ligated. The pulmonary end of the graft was resected from the right pulmonary artery and the defect was repaired with a small pericardial patch. The main pulmonary artery was incised and pulmonary valvotomy together with transannular patch with an autologous pericardium were performed to relieve pulmonary stenosis

https://doi.org/10.1017/S1047951112001308 Published online by Cambridge University Press

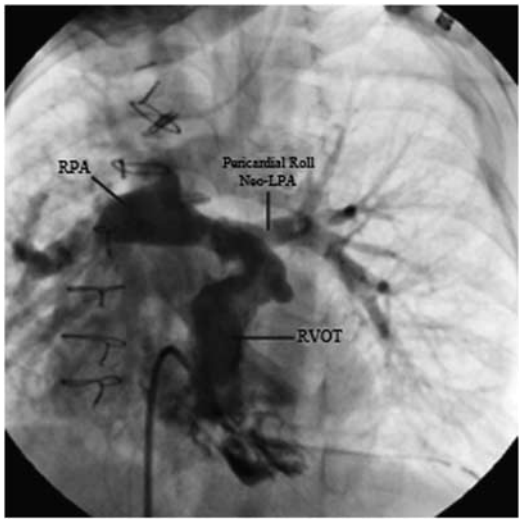

Figure 4.
Control angiography right ventricular injection indicating reconstructed right ventricular outflow tract, pulmonary arteries as well as pericardial roll reconstructed left pulmonary artery. LPA = left pulmonary artery; RPA = right pulmonary artery; RVOT = right ventricular outflow tract.

(Brock procedure). Weaning off the cardiopulmonary bypass was possible with moderate dose inotropic support (7 micrograms per kilogram per minute of dopamine, 7 micrograms per kilogram per minute of dobutamin, and 0.05 micrograms per kilogram per minute of adrenalin). Mean 23 millimetres of mercury transpulmonary gradient and 73% oxygen saturation with 20% inspiratory oxygen were measured. Total circulatory arrest, cross-clamp, and cardiopulmonary bypass times were 18, 35, and 59 minutes, respectively. She was taken to the intensive care unit and extubated there the next day.

The post-operative course was uneventful. Control angiography showed successfully reconstructed right ventricular outflow tract and pulmonary arteries (Fig 4). She was discharged from the hospital on the 10th day with 83% room air oxygen saturation and scheduled for a corrective surgery.

Discussion

Double outlet right ventricle is a congenital cardiac malformation, which ranges between tetralogy of Fallot and transposition of great arteries. The incidence of the pathology is about 1% among congenital heart defects.[2] It is a type of ventriculoarterial connection abnormality in which both the pulmonary artery and the aorta arise from the right ventricle.[4] The pathology differs from that of tetralogy of Fallot, with more than 50% aortic override and loss of the aortic-mitral fibrous continuity. On the other hand, in case of transposition of the great arteries, there is pulmonic-mitral continuity. There are bilateral muscular coni that carry the aorta and the pulmonary artery anteriorly and superiorly in the double outlet right ventricle pathology.[2,4]

Unilateral absent pulmonary artery is a very rare congenital malformation with an incidence less than 1%.[5,6] It may accompany other congenital cardiac defects such as ventricular septal defect, coarctation of the aorta, subvalvular aortic stenosis, transposition of the great arteries, pulmonary stenosis,[6] and the most common being tetralogy of Fallot.[7,8] More frequently, the left pulmonary artery is absent,[5–8] although absent right pulmonary artery has also been reported.[5,9] Interestingly, in the literature there is only one patient reported to have double outlet right ventricle and absent left pulmonary artery.[3]

There are several theories for the unilateral absence of pulmonary arteries. During the involution of the right or the left sixth arch, the corresponding right or the left pulmonary artery may also disappear[9] or there may be an abnormal division of the truncus; that is, the dorsal shift of the ridges may cause agenesis of the corresponding pulmonary artery.[9] The latter is called the ontogenetic theory and may explain the absence of the right pulmonary artery as an isolated defect and the left pulmonary artery being associated with other congenital malformations.[8]

The symptoms of patients with unilateral absent pulmonary artery with additional cardiac malformations are usually related with the cardiac defect. The pulmonary artery absence adds over them. Rarely, patients may be asymptomatic or sometimes may present with symptoms related to pulmonary hypertension and congestive cardiac failure such as dyspnoea, fatigue, or haemoptysis.[10] In our case, the patient had cyanosis and associated symptoms.

Conventional radiologic imaging tools readily facilitate the diagnosis. Chest roentgenogram gives an idea about the pulmonary status. Echocardiography is very helpful and cardiac catheterisation is still the gold standard for the diagnosis and surgery planning for such challenging cases. Pulmonary venous wedge angiography is useful to investigate the presence of a pulmonary artery at the hilum of the lung and intrapulmonary vessels.[10] Magnetic resonance imaging and computerised tomography are also valuable tools.

Surgical treatment of patients with single pulmonary artery with associated cardiac defects is challenging. Both staged approach and correction in different series of tetralogy of Fallot and unilateral absent pulmonary artery have considerable mortality and morbidity rates.[7,8] Patients with pulmonary hypertension and congestive cardiac failure do not

https://doi.org/10.1017/S1047951112001308 Published online by Cambridge University Press

benefit from surgery except transplantation. In the report of Kremer et al,[3] authors preferred single-stage correction in their patient; however, we preferred a staged approach in our patient owing to the low pulmonary indices.

In conclusion, double outlet right ventricle with unilateral absent left pulmonary artery has been reported as a single case report only once in the literature.[3] To the best of our knowledge, this is the second patient with a similar pathology in whom a palliative procedure was preferred and the post-operative course had been complex.

References

1. Atay Y, Iyem H, Yagdi T, Levent E, Ozyurek R, Alayunt EA. Double outlet right ventricle. Turkish J Thorac Cardiovasc Surg 2004; 12: 64–69.
2. Männer J, Seidl W, Steding G. Embryological observations on the morphogenesis of double-outlet right ventricle with subaortic ventricular septal defect and normal arrangement of the great arteries. Thorac Cardiovasc Surg 1995; 43: 307–312.
3. Kremer S, Fayemi AB, Fish BG, Rothe WA, Spindola-Franco H. Congenital absence of the left pulmonary artery associated with double-outlet right ventricle. AJR Am J Roentgenol 1992; 158: 1309–1311.
4. Walters HL III, Mavroudis C, Tchervenkov CI, Jacobs JP, Lacour-Gayet F, Jacobs ML. Congenital heart surgery nomenclature and database project: double outlet right ventricle. Ann Thorac Surg 2000; 69 (Suppl 4): S249–S263.
5. Kucera V, Fiser S, Tuma S, Hucin B. Unilateral absence of pulmonary artery: a report on 19 selected clinical cases. Thorac Cardiovasc Surg 1982; 30: 152–158.
6. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. Chest 2002; 122: 1471–1477.
7. Zhang GC, Wang ZW, Zhang RF, Zhu HY, Yi DH. Surgical repair of patients with tetralogy of Fallot and unilateral absence of pulmonary artery. Ann Thorac Surg 1997; 64: 1150–1153.
8. Bockeria LA, Podzolkov VP, Makhachev OA, et al. Surgical correction of tetralogy of Fallot with unilateral absence of pulmonary artery. Ann Thorac Surg 2007; 83: 613–618.
9. Ugurlucan M, Arslan AH, Yildiz Y, Ay S, Besikci RT, Cicek S. Tetralogy of Fallot with unilateral absent pulmonary artery. Cardiol Young 2012: 1–8.
10. Halit V, Olguntürk R, Erer D, Kula S. Tetralogy of Fallot and absence of left pulmonary artery. Thorac Cardiovasc Surg 2008; 56: 430–432.