Cardiology in the Young

cambridge.org/cty

A situs solitus transposition of great arteries with obstructed sub-diaphragmatic totally anomalous pulmonary venous connection: a rare case treated with anatomical repair

Giovanni Meliota, Gabriele Scalzo and Ugo Vairo

Department of Paediatric Cardiology, Giovanni XXIII Paediatric Hospital, Bari, Italy

Brief Report

Cite this article: Meliota G, Scalzo G, and Vairo U (2019) A situs solitus transposition of great arteries with obstructed sub-diaphragmatic totally anomalous pulmonary venous connection: a rare case treated with anatomical repair. *Cardiology in the Young* **29**: 1536–1538. doi: 10.1017/S1047951119002415

Received: 1 August 2019
Revised: 31 August 2019
Accepted: 10 September 2019
First published online: 30 October 2019

Keywords:
Transposition of great arteries; totally anomalous pulmonary venous connection; arterial switch operation

Author for correspondence:
Giovanni Meliota, Paediatric Cardiology, Giovanni XXIII Paediatric Hospital, Via Amendola, 207 70126 – Bari, Italy. Tel: +39 080 5596698; Fax: +39 080 5596700; E-mail: giovanni.meliota@gmail.com

Abstract

Transposition of the great arteries combined with totally anomalous pulmonary venous connection is extremely rare outside of heterotaxy syndrome. Most reported cases have been treated by a modified atrial switch operation. We report the successful treatment of a neonate with this rare association, repaired by arterial switch operation and connection of the pulmonary venous return to the left atrium.

Transposition of the great arteries combined with totally anomalous pulmonary venous connection is extremely rare outside of heterotaxy syndrome. Since the first description in 1964,[1] few cases have been reported,[2–11] most have been treated by an atrial switch operation. We report the successful treatment of a neonate with transposition of the great arteries, intact ventricular septum and obstructed sub-diaphragmatic totally anomalous pulmonary venous connection, repaired by arterial switch operation and connection of the pulmonary venous return to the left atrium.

Case presentation

A female neonate was delivered at term via urgent caesarean section due to reduced foetal movements and podalic presentation. The birth weight was 2850 g and the Apgar scores were 8 and 9. Central cyanosis was noted within a few hours that was unresponsive to oxygen administration but partially resolved with prostaglandin infusion.

The infant was irritable and tachypneic but not dysmorphic. The heart rate was 160 beats per minute, oxygen saturation on room air was 85% and blood pressure was 80/50 mmHg and equal in all extremities. S2 was loud, and a soft blowing ejection murmur was audible at the upper left sternal border. The exam was otherwise normal.

The electrocardiogram (EKG) demonstrated sinus rhythm with right ventricular hypertrophy. The initial echocardiographic diagnosis was levocardia, transposition of the great arteries {S,D,D} (D-transposition of the great arteries) (Fig 1a), large secundum atrial septal defect with right-to-left shunt and a patent arterial duct (Fig 1b). The right heart chambers appeared dilated.

Given the unrestrictive atrial communication, balloon atrial septostomy was considered unnecessary. The patient was scheduled for an arterial switch operation.

Prompted by the right-to-left atrial shunt, a repeat echocardiogram documented the absence of pulmonary venous return in the left atrium. A large vascular channel was interposed between the descending aorta and inferior caval vein, with phasic venous flow directed towards the abdomen consistent with sub-diaphragmatic totally anomalous pulmonary venous connection (Fig 1c). The venous channel showed a connection to the portal sinus (Fig 1d) with a restrictive venous duct characterised by a high-velocity, continuous, and non-phasic venous flow profile, consistent with an obstructed anastomosis.

Given the extreme rarity of this association, it was decided to further investigate the anatomy, to best plan the surgical repair. Cardiac catheterisation and angiography confirmed the diagnosis and demonstrated usual coronary anatomy for D-transposition of the great arteries (Fig 2).

The surgical repair consisted of anastomosis of the pulmonary venous confluence to the left atrium, arterial switch operation and atrial septal defect closure with a 3.5-mm-fenestrated bovine pericardial patch. Inhaled nitric oxide administration was started due to pulmonary artery hypertension and sternal closure was delayed.

Inhaled nitric oxide therapy was stopped as pulmonary artery pressure normalised, and the chest was closed on the 4th post-operative day. The patient was extubated the following day. Recovery was complicated by a right chylothorax that resolved with thoracic drainage, octreotide

CAMBRIDGE UNIVERSITY PRESS

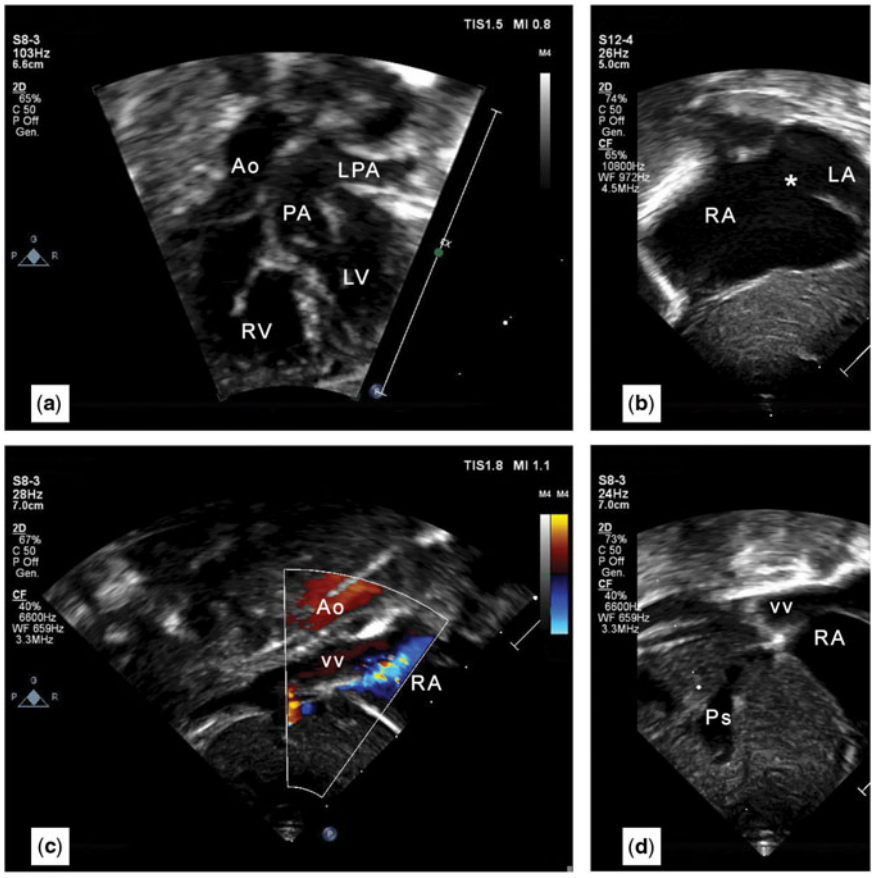

Figure 1. Echocardiographic 2D images. (***a***) In this subxiphoid view, D-transposition of the great arteries with intact ventricular septum is clearly demonstrated. (***b***) There is a large secundum atrial septal defect (*asterisk*) with right-to-left shunt. RA is enlarged. (***c***) Subxiphoid short-axis imaging with colour Doppler mapping shows a large vertical vein coursing inferiorly from behind the heart through the diaphragm; this is anterior to the aorta (Ao). Flow is towards the probe (red) in both vessels. (***d***) Colour aliasing indicates an obstruction of the vertical vein at the site of entry into the PS, which is dilated. Ao = aorta; LPA = left pulmonary artery; LV = left ventricle; PA = pulmonary artery; PS = portal sinus; RA = right atrium; RV = right ventricle; VV = vertical vein.

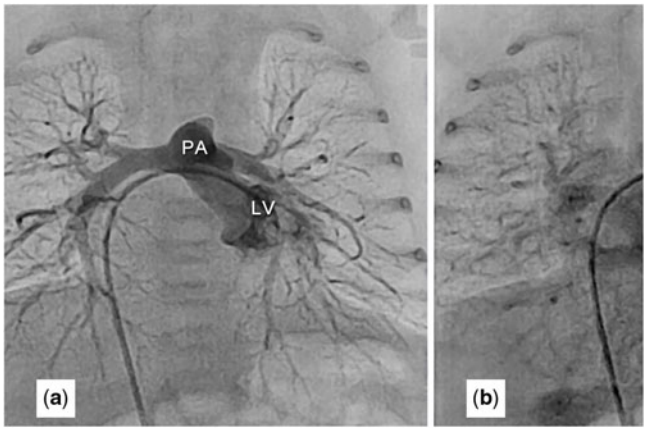

Figure 2. Cardiac catheterisation. Catheter course: femoral vein – inferior vena cava – right atrium – left atrium – left ventricle. (***a***) The left ventricular angiogram shows an intact ventricular septum and the PA arising from the LV, thus demonstrating the D-transposition of the great arteries. (***b***) Levophase of the LV angiogram showing all pulmonary veins draining into a large VV, with an inferior course through the diaphragm and a restrictive connection to the portal sinus. LV = left ventricle; PA = pulmonary artery; VV = vertical vein

infusion, intravenous furosemide and a medium chain triglyceride diet. The patient was discharged on the 14th post-operative day in good clinical condition.

The patient is now doing well at 1-year follow-up. The last echocardiography documented mild neopulmonary valve stenosis with preserved biventricular systolic function, normal chamber dimensions and unobstructed pulmonary veins.

Discussion

D-transposition of the great arteries with totally anomalous pulmonary venous connection is an extremely uncommon association, except as part of heterotaxy syndrome.[12] To the best of our knowledge, in the setting of a definite visceroatrial situs (solitus or inversus), only 11 in vivo cases of D-transposition of the great arteries with totally anomalous pulmonary venous connection have been reported (Table 1). Among post-mortem[12] and in vivo reports, sub-diaphragmatic drainage was the rarest form of totally anomalous pulmonary venous connection, with only three cases before ours.[5,8,11] Mykychak described the only case with the same anatomy as our report (intact ventricular septum and no other associated CHDs).[11]

As in Lopes et al,[8] our first echocardiogram did not detect the totally anomalous pulmonary venous connection. While in that report totally anomalous pulmonary venous connection was diagnosed during cardiac catheterisation, in our case a repeat echocardiogram revealed the totally anomalous pulmonary venous connection before catheterisation. The clues to the diagnosis were an unusual aliased flow pattern below the diaphragm and the unidirectional right-to-left atrial shunt with right heart dilation.

These two defects together partly correct the abnormal haemodynamics caused by each one alone. Totally anomalous pulmonary venous connection directs pulmonary venous blood to the right

https://doi.org/10.1017/S1047951119002415 Published online by Cambridge University Press

Table 1. Transposition of the great arteries and totally anomalous pulmonary venous connection: in vivo cases reported in the literature

Year	Author	Age/Sex	Type of TAPVC	Associated CHD	Diagnosis	Surgery
1964	Whitaker[1]	9 year/F	Supracardiac	None	Cath	None
1972	Sapsford[2]	16 year/F	Supracardiac	None		Atrial (Mustard)
1981	Barbero-Marcial[3]	8 months/M	Supracardiac	None	Cath	Atrial (Senning)
1990	Amodeo[4]	2 months/F	Cardiac (RA)	VSD	Echo, Cath	Atrial (Mustard)
1990	Thies[5]	3 days/M	Sub-diaphragmatic	VSD, cor triatriatum	Echo, Cath	Atrial (Mustard)
1994	Ueda[6]	1 year/M	Supracardiac	None	Cath	Atrial (Senning)
1994	Gontijo[7]	11 months/F	Cardiac (CS)	None	Intraop	Atrial (Mustard)
2001	Lopes[8]	22 days/M	Sub-diaphragmatic	VSD	Cath	ASO
2002	Raff[9]	13 years/F	Supracardiac	Severe TR	Unknown	Atrial (Senning)
2016	Salve[10]	6 days/M	Cardiac (CS)	LJAA	Echo	ASO
2017	Mykychak[11]	Prenatal/M	Sub-diaphragmatic	None	Echo, MRI	ASO
2019	Meliota	6 days/F	Sub-diaphagmatic	None	Echo, Cath	ASO

ASO = arterial switch operation; Cath = cardiac catheterisation; CS = coronary sinus; Eco = echocardiography; Intraop = intraoperative; LJAA = left juxtaposition of atrial appendages; RA = right atrium; TAPVC = total anomalous pulmonary venous connection; TR = tricuspid regurgitation; VSD = ventricular septal defect

heart from which it is pumped to the aorta due to D-transposition of the great arteries, increasing systemic arterial saturation. This combination can produce fewer symptoms, milder cyanosis, and result in later diagnosis than the isolated defects.

Since the first successful repair in 1972,[2] most reported cases have been treated by a modified atrial switch operation (Table 1), at least in part due to late diagnosis as discussed above. Some have even proposed that D-transposition of the great arteries with totally anomalous pulmonary venous connection precludes neonatal arterial switch operation because the left ventricle is not prepared to handle the systemic circulation, because of reduced left heart flow during foetal life.[7] This seems unlikely because pulmonary blood flow in utero accounts for only about 10% of combined ventricular output and three patients diagnosed as neonates underwent an arterial switch operation with satisfactory results.

Conclusion

Although extremely rare, transposition of the great arteries can be associated with anomalous pulmonary venous drainage even with normal atrial situs. A systematic, segmental approach to echocardiographic examination is essential to avoid overlooking important diagnoses. In D-transposition of the great arteries, special attention should be given to unusual indirect findings, such as a continuous right-to-left atrial shunt and disproportionately dilated right chambers, as they can be hints for an associated major anomaly. If diagnosed within the first weeks of life, arterial switch operation should be preferred over an atrial switch operation, as it avoids late complications related to a systemic right ventricle.

Acknowledgements. We would like to acknowledge Prof. Stephen P. Sanders (Boston Children's Hospital, Boston, MA, USA) for his review of the report and valuable suggestions.

Financial Support. This work received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest. None.

Ethical Standards. No human or animal experimentation was used.

References

1. Whitaker W, Watson DA, Keates PG. Total anomalous pulmonary venous drainage into the left innominate vein associated with transposition of the great vessels. Circulation 1964; 30: 918–922.
2. Sapsford RN, Aberdeen E, Watson DA. Transposed great arteries combined with totally anomalous pulmonary veins. Report of a successful repair. J Thorac Cardiovasc Surg 1972; 63: 360–366.
3. Barbero-Marcial M, Verginelli G, Vila J, Zerbini EJ. Transposition of the great arteries associated with total anomalous pulmonary venous connection: a surgical approach. Ann Thorac Surg 1984; 37: 92–94.
4. Amodeo A, Corno A, Marino B, Carta MG, Marcelletti C. Combined repair of transposed great arteries and total anomalous pulmonary venous connection. Ann Thorac Surg 1990; 50: 820–821.
5. Thies WR, Matthies W, Minami K, Pott U, Meyer H, Körfer R. Surgical repair and postoperative course of an infant with infracardiac total anomalous pulmonary venous connection, cor triatriatum sinistrum and transposition of the great arteries. Eur J Cardiothorac Surg 1990; 4: 45–47.
6. Ueda Y, Miki S, Okita Y, et al. Transposition of the great arteries associated with total anomalous pulmonary venous return. Ann Thorac Surg 1994; 57: 470–472.
7. Gontijo B, Fantini F, Barbosa M, Gomes MV, Gutierrez C, Vrandecic M. Surgical repair of transposition of great arteries and total anomalous pulmonary venous return to the coronary sinus (TGA with TAPVR). Eur J Cardiothorac Surg 1994; 8: 391–392.
8. Lopes LM, Tavares GMP, Mailho FL, Almeida VD, Mangione JA. Echocardiographic diagnosis of transposition of the great arteries associated with anomalous pulmonary venous connection. Arq Bras Cardiol 2001; 77: 63–68.
9. Raff GW, Geiss DM, Shah JJ, Bond LM, Carroll JA. Repair of transposition of the great arteries with total anomalous pulmonary venous return. Ann Thorac Surg 2002; 73: 655–657.
10. Salve GG, Jain SA, Dalvi BV, Shivaprakash K. Transposition of the great arteries with total anomalous pulmonary venous connection. Ann Thorac Surg 2017; 103: e349–e351.
11. Mykychak Y, Fedevych O, Maksymenko A, Yemets I. Simultaneous arterial switch and totally anomalous pulmonary venous connection repair in a 5-hour-old child, complicated by pulmonary venous stenosis. Interact Cardiovasc Thorac Surg 2017; 24: 809–810.
12. Delisle G, Ando M, Calder AL et al. Total anomalous pulmonary venous connection: report of 93 autopsied cases with emphasis on diagnostic and surgical considerations. Am Heart J 1976; 91: 99–122.

https://doi.org/10.1017/S1047951119002415 Published online by Cambridge University Press