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CASE REPORT

Cor Triatriatum With Partial Anomalous Pulmonary Venous Return: A Rare Case of Parallel Obstruction and Successful Staged Treatment

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Abstract Cor triatriatum sinister is an uncommon congenital cardiac defect that has rarely been described in association with left-sided partial anomalous pulmonary venous return. We present a case of such rare anatomy with multilevel obstruction that presented in infancy as cardiogenic shock. The patient underwent staged treatment with extracorporeal membrane oxygenation stabilization, catheter-based balloon dilatation of the cor triatriatum and atrial septostomy, followed by definitive surgical repair, with excellent result.

Keywords Cor triatriatum · Partial anomalous pulmonary venous return · Pulmonary venous obstruction · Cardiac catheterization · Cardiac surgery

Introduction

Cor triatriatum sinister (CTS) is a rare congenital cardiac defect with an incidence of 0.2–0.4% at autopsy [6]. First described in 1868, CTS occurs when the embryologic common pulmonary vein fails to incorporate into the back of the left atrium, resulting in a fibromuscular intra-atrial membrane. The communication between the proximal CTS chamber and the distal (true) left atrium may be severely stenotic, thereby compromising inflow to the left ventricle. Many variations of CTS exist, including anomalous connection of the cor chamber to the right atrium or systemic veins. In subtotal CTS, the cor receives one to three of the pulmonary veins [5]. Most patients present during infancy with dyspnea, failure to thrive, and frequent respiratory infections [8]. Cor triatriatum has been reported with right-sided partial anomalous pulmonary venous return (PAPVR) in multiple cases. In contrast, PAPVR of the left pulmonary veins with CTS is a less described association [1–4, 7]. The combination of an obstructive CTS and left-sided PAPVR with a restrictive atrial communication is exceedingly rare. We describe the use of elective short-term extracorporeal membrane oxygenation (ECMO) support for stabilization in the context of such parallel obstruction of left-ventricular inflow.

Case Report

A 3-month-old full-term baby girl presented to the emergency department with several days of cough, irritability, and progressive lethargy. On physical examination, she had moderate respiratory distress, poor perfusion, and hepatomegaly. Although tachycardic, her blood pressure was maintained, but her pulse oximetry was 85% on room air. Laboratory evaluation was notable for severe lactic acidosis and evidence of a urinary tract infection. Chest X-ray showed significant cardiomegaly and symmetrical pulmonary venous congestion. A stat echocardiogram performed in the emergency department demonstrated an obstructed CTS membrane in the left atrium (with peak gradient of 23 mm Hg across the cor orifice) as well as PAPVR, with the left pulmonary veins draining to an ascending vertical vein to the left innominate vein. There was right-to-left atrial level

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shunting through a patent foramen ovale (PFO) into the distal left atrium. The right pulmonary veins drained into the obstructive cor chamber (Fig. 1). Pulmonary artery pressure was near-systemic with a TR maximal gradient of 51 mm Hg, and right-ventricular function was severely decreased (see Supplementary Movie Clips 1 and 2).

The patient was intubated and resuscitated with volume, bicarbonate, and dopamine infusion. A decision to perform a staged treatment was made. Due to persistent lactic acidosis and the understanding that she would require a palliative catheter-based intervention before definitive surgical repair, she was electively deployed on a veno-arterial ECMO through a neck approach. On hospital day 2, after initial stabilization, she underwent dynamic balloon septostomy of the atrial septum as well as high-pressure balloon dilatation of the CTS membrane using a 13-mm balloon (see Supplementary Movie Clip 3). The gradient over the cor membrane orifice was decreased from 20 to 1 mm Hg (Fig. 2 and Supplementary Movie Clip 4). There was no significant obstruction of the left pulmonary vein drainage to the innominate vein. The catheterization as well tolerated.

By hospital day 3, the patient's liver and renal indices had completely normalized. The patient was decannulated from ECMO on hospital day 6 and extubated the next day. On hospital day 10, she underwent surgical resection of the CTS membrane, repair of PAPVR by division of the vertical vein from the innominate vein, end-to-side anastamosis of the vertical vein to the left atrium, and pericardial patch closure of the atrial septal defect. She had an uncomplicated postoperative course, and was discharged home on hospital day 16. Her postoperative echocardiogram showed no residual disease. After 9 months of follow-up, she remains asymptomatic and is thriving with no gradient over the ascending vein-left atrium anastomosis, normal bi-ventricular function and normal pulmonary pressure on echocardiography.

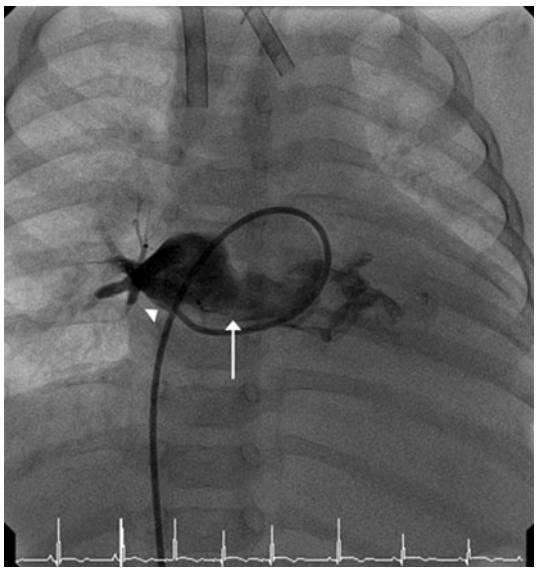

Fig. 2 Postintervention angiographic view of the cor triatriatum chamber with good blood egress to the true distal left atrium through the dilated cor orifice (*arrow*). The *arrowhead* points to the atrial septum plane

Discussion

We present an infant with a rare combination of severely obstructed subtotal CTS that drained the right pulmonary veins with associated partial anomalous connection of the left pulmonary veins to the innominate vein. Because the PFO was also severely restrictive, the egress of blood from

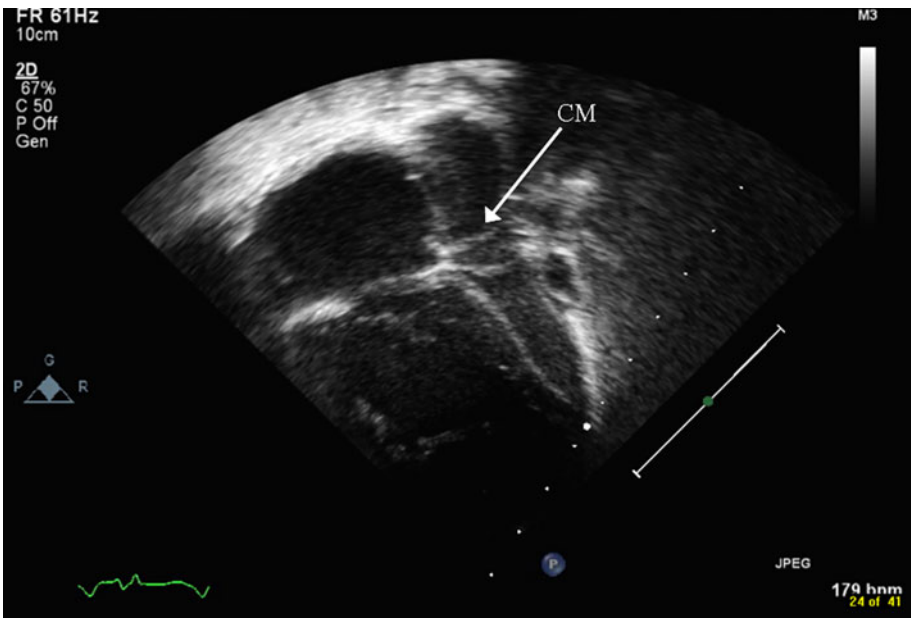

Fig. 1 Transthoracic apical four-chamber echocardiographic view of the cardiac anatomy. *CM* cor triatriatum membrane

the pulmonary veins into the systemic ventricle was severely impaired, manifesting as cardiogenic shock.

The first successful surgical repair of CTS associated with left PAPVR was reported in 1977 by Jennings et al. [7] in a 10-week-old infant who had an obstructed CTS that drained the right pulmonary veins, whereas the left pulmonary veins drained in an unobstructed manner to the right atrium. Throughout the years rare cases of left PAPVR have been published with association of CTS [2, 4]. Krasemann et al. [8] reviewed the literature on CTS and described ten cases in 2007. Most of their patients had associated cardiac anomalies, including atrial septal defect, ventricular septal defect, or pulmonic stenosis. Only one patient had associated PAPVR. The majority of the patients were symptomatic with tachypnea and failure to thrive, presumably due to obstruction of the cor membrane orifice.

Due to the poor condition of our patient at presentation and the intercurrent infection, we elected to perform "staged treatment." After delineating the anatomy by echocardiography, we believed it would be safer to electively deploy the patient on ECMO support. ECMO has rarely been used for anomalous venous return, and this semiurgent procedure not only enabled initial stabilization and prevention of further end-organ insult but also facilitated the catheter-based intervention in a more secure fashion. Catheterization confirmed the anatomy, documented the severity of the obstruction, and relieved the obstruction of the bilateral pulmonary venous drainage. The resultant increase in cardiac output enabled the patient to be quickly weaned off ECMO and mechanical ventilatory support. Before surgical repair 3 days later, the patient's condition had completely stabilized. The operative procedure was the last stage of the treatment plan, and her recovery was fast and uneventful.

Conclusion

To our knowledge, this is the first reported case of obstructed CTS with associated obstructed PAPVR at the atrial level. The infant presented in cardiogenic shock, which was probably precipitated by an inability to increase cardiac output in the context of an intercurrent urinary tract infection. The staged-treatment approach and the novel use of ECMO for short-term stabilization for a palliative catheterization procedure allowed for final surgical repair when the patient was in the optimal medical condition.

References

1. Camilleri L, Lusson JR, Picarelli D, d'Agrosa MC, Peycelon B, de Riberolles C (1991) Association of cor triatriatum and obstruction of abnormal left pulmonary venous drainage. Apropos of a case. Arch Mal Coeur Vaiss 84(3):409–412
2. Cooley DA, Murphy MC (1990) Cor triatriatum and anomalous pulmonary venous return: An unusual case. Tex Heart Inst J 17(2):118–121
3. Fujimatsu T, Tsunemoto M, Shimada M, Ohta Y, Endo S, Ishizawa A et al (1990) Subtotal cor triatrium with left partial anomalous pulmonary venous connection—Successful correction in an infant. Rinsho Kyobu Geka 10(1):76–80
4. Geggel RL, Fulton DR, Chernoff HL, Cleveland R, Hougen TJ (1987) Cor triatriatum associated with partial anomalous pulmonary venous connection to the coronary sinus: echocardiographic and angiocardiographic features. Pediatr Cardiol 8(4):279–283
5. Geva T, Van Praagh S (1995) Abnormal pulmonary venous connections. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF (eds) Heart disease in infants, children, and adolescents, 5th edn. Williams & Wilkins, Baltimore, pp 761–792
6. Jegier W, Gibbons GJ, Wiglesworth FW (1963) Cortriatriatum: clinical, hemodynamic and pathological studies surgical correction in early life. Pediatrics 31:255–267
7. Jennings RB Jr, Innes BJ (1977) Subtotal cor triatriatum with left partial anomalous pulmonary venous return. Successful surgical repair in an infant. J Thorac Cardiovasc Surg 74(3):461–466
8. Krasemann Z, Scheld HH, Tjan TD, Krasemann T (2007) Cor triatriatum: short review of the literature upon ten new cases. Herz 32(6):506–510