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Journal of Cardiac Disorders and Therapy

CASE REPORT

ISSN: 2637-465X

Late Diagnosis of Total Anomalous Connection of Pulmonary Veins during the Puerperium

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Citation: Pumacayo-Cárdenas S, Quea-Pinto E (2019) Late Diagnosis of Total Anomalous Connection of Pulmonary Veins during the Puerperium. J Card Disord Therapy 2: 101

Article history: Received: 25 May 2019, **Accepted:** 05 July 2019, **Published:** 08 July 2019

Abstract

The total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly. We present a case of a 36-year-old woman, in the postpartum period during the physical examination; there was heart murmur and beating in hands and feet. Cardiac magnetic resonance reported supracardiac TAPVC and large atrial septal defect. The survival of these patients is very uncommon in adults and even worse during pregnancy.

Keywords: Anomalous Systemic Venous Drainage; Postpartum Period; Adult; Phase-Contrast MRI

Introduction

The total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly, which corresponds to approximately 2% of all congenital heart defects [1,2]. The majority of patients present in the neonatal period with decompensated heart failure due to severe pulmonary hypertension or obstruction of pulmonary venous return [3,4]. The natural history of the disease shows that 20% survive until the first year of life without treatment [5]. Exceptionally, the symptoms may appear later in life or progressively, depending on some factors, which will be discussed later [6], the case we present is of a woman who is diagnosed with congenital heart disease during the puerperium. In some series, maternal mortality of patients with pulmonary hypertension, during pregnancy and the puerperium is very high.

Presentation of the case

A 36-year-old female patient, from 13 years of age, began with dyspnea at great exertion and cyanosis of the lips. At 35 years of age, during her first pregnancy, she decline in her functional class. She went to a health care center for the first time to attend her delivery, which was eutocic, without hemodynamic complications. The product was a newborn, full term, with adequate weight for gestational age, healthy. The hospital discharge was usually the second postpartum day. The findings in the physical examination were cyanosis, digital clubbing, without signs of respiratory distress (oxygen saturation 85%, FiO_2 24%), the hyperdynamic precordium, cardiac auscultation revealed a regular heart rhythm, a wide and fixed division of the second heart sound with accentuation of the pulmonary component and holosystolic murmur 4/6 in the pulmonic area irradiated to the neck. Liver palpable 4 cm below the costal margin. The peripheral pulses were symmetrical and there was no peripheral edema. The chest x-ray revealed cardiomegaly, and cardiac silhouette in the shape of a "snowman" (Figure 1A). Echocardiography was not optimal, found dilation of right cavities, atrial septal defect (ASD) of 15 mm and systolic pulmonary pressure of 70 mmHg. Twenty days postpartum, the patient comes to our institution, where the magnetic resonance imaging (MRI) shows the presence of total anomalous connection of pulmonary veins supracardiac variety. The confluence of the four pulmonary veins was wide (diameter 36 x 25 mm) and was connected to a dilated vertical vein, which in turn drained into a dilated innominate vein (at the mouth its diameter measured 32 x 20 mm), without evidence of obstruction (Figure 1 By 2). Right atrium measured 23 cm^2 / m^2, left atrium 7.5 cm^2/m^2. The ASD was superior venous sinus type of 25 x 20mm, shunt from right to left and high pulmonary flow (Qp:Qs 2.9). Dilated right ventricle, systolic septal flattening, ejection fraction 48% and moderate tricuspid regurgitation; left ventricle with preserved ejection fraction. Trunk and branches of the pulmonary artery dilated. Presence of late gadolinium enhancement intramyocardial (Figure 2B).

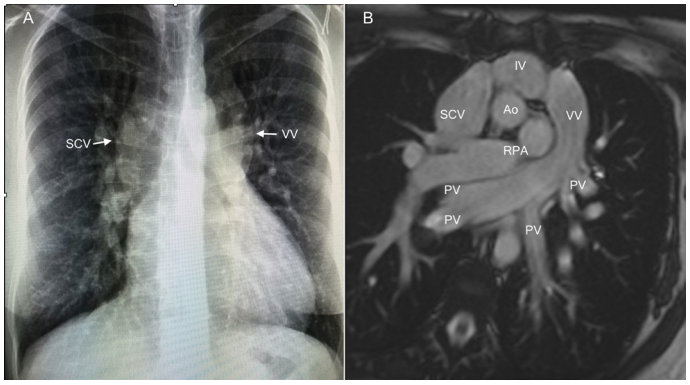
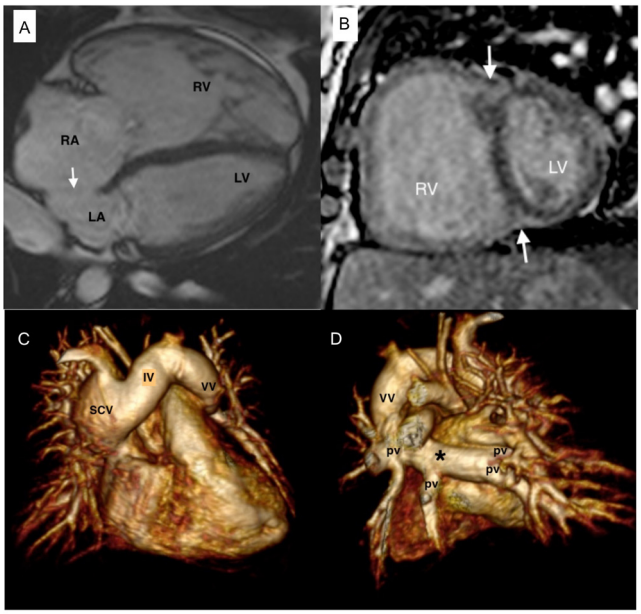


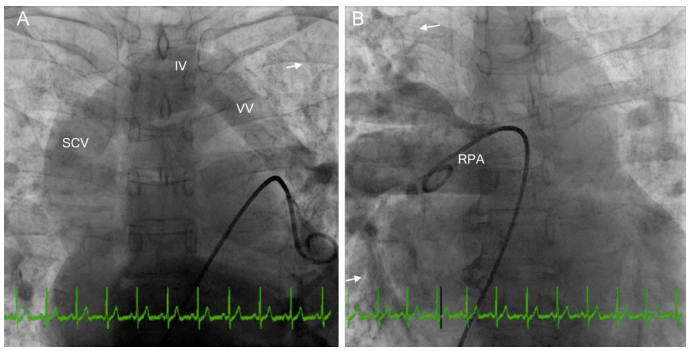
Figure 1: Chest X-ray (A) Configuration of the heart and the superior mediastinal borders resembling a "snowman"; **(B)** Echo gradient acquisition shows the pulmonary veins (PV) drain into an enlarged vertical vein (VV) which in turn drains into the innominate (IV) vein and then to the superior vena cava (arrow); Right pulmonary artery (RPA)



VV: Vertical Vein; **IV:** Innominate Vein; **RA:** Right Atrium; **SVC:** Superior Vena Cava

Figure 2: (A) Axial echo gradient acquisition shows four chamber view, showed that the right atrium and ventricle were significantly enlarged, the left atrium was small without pulmonary vein entrance. Note a large atrial septal defect (arrow); **(B)** Arrows point at zones of late gadolinium enhancement pattern of mid-wall fibrosis in sites of union of the left ventricle (LV) with the right and in the inferior wall of the RV; **(C)** The 3-D MRI image (anterior view) y; **(D)** Posterior view, showed five pulmonary veins (pv) entered into a pulmonary venous sinus (*) connected through the vertical vein to innominate vein which entered into a significantly enlarged superior vena cava

Cardiac catheterization (Figure 3) reported pulmonary arterial hypertension, due to volume overload, Qp:Qs 3; even without involvement of the pulmonary microvasculature: mean of the pulmonary pressure 60 mmHg, pulmonary vascular resistance (PVR) 2.5 U Wood/m^2, the ratio of pulmonary vascular resistance and systemic (PVR:SVR) was 0.1. The systemic pressure (SP) was 115mmHg; the pulmonary pressure was less than 2/3 of SP. With these hemodynamic parameters, it was decided to perform the respective total surgical repair.



VV: Vertical Vein; **IV:** Innominate Vein; **RA:** Right Atrium; **SVC:** Superior Vena Cava

Figure 3: Cardiac Catheterization (A) Angiography in the left pulmonary artery (LPA), showed the absence of venous catheter obstruction (adequate contrast step in the drainage of the VV to the IV vein and from this to the SCV; **(B)** Angiography in the right pulmonary artery (RPA), showed significant dilatation to the lobar and segmental branches, the distal vasculature is preserved (arrows)

Postoperative evolution was favorable, did not present a pulmonary hypertension crisis, or arrhythmias, with support of inotropes during the first two days. As of the third postoperative day, the systolic pulmonary pressure obtained by echocardiography was 35 mmHg, the biventricular systolic function remained normal, without residual defects. She was discharged 10 days after surgery, without complications.

Discussion

Due to the large variations in anatomy and hemodynamics, TAPVC demonstrates a broad spectrum of clinical presentations ranging from the absence of symptoms to severe hypoxemia [8-11]. To survive to adulthood, there should be no obstruction between the pulmonary venous catheter, the systemic vein and the right atrium, an adequate size of the ASD, adequate oxygenation of the arterial blood and exclude the presence of severe pulmonary hypertension. Jian, *et al.* [11] in their study describes the average diameter of the pulmonary venous confluence (25mm), the average size of the atrial septal defect (31mm) and the non-restrictive drainage site can provide conditions compatible with long-term survival term. In the present case, similar conditions were observed.

The thorough evaluation of these patients must include right cardiac catheterization, for the measurement of pulmonary pressure, PVR. Criteria for the closure of the short circuit have been proposed based on the baseline RVP <4 U Wood / m^2ASC. Additional criteria are the type of defect, the age, the ratio PVR: SVR and the quotient Qp: Qs. There are no prospective data on the usefulness of the vasoreactivity tests, the closure tests or the lung biopsy for the evaluation of operability; however, its use will depend on the treating doctor [12].

In patients with PH, characterized by having fixed and elevated RVP, there is great difficulty in managing high cardiac output during pregnancy and its increase during delivery, which leads to higher mortality. Clinical deterioration appears more frequently in the second trimester of pregnancy, corresponding to a 40% increase in cardiac output [12]. The diagnosis of this case is very rare in the adult, due to the high infant mortality when the corrective treatment is not carried out. The presence of a broad atrial septal defect, which allowed blood flow from right to left, the absence of obstruction of the venous system and the preservation of the pulmonary microvasculature prevented a fatal event during the development of this patient's life, in this way could be a candidate for corrective treatment.

Conclusions

The diagnosis of TAPVC is infrequent in adulthood. The case presented is exceptional, as it is an adult woman and in the postpartum period. Survival in these cases will depend on the absence of pulmonary venous obstruction, presence of large intracardiac septal defects that allow right to left shunts. However, the presence of severe pulmonary hypertension and damage to the pulmonary vasculature is common and may limit the corrective treatment. MRI and catheterization are suitable diagnostic methods, for use in preoperative evaluation.

References

1. Sulaiman S, Nair RG (2017) Anomalous left atrial drainage of the right superior caval vein with partial anomalous pulmonary venous return. Cardiol Young 27: 1216-8.

2. Ahmad N, Sell JE (1993) Closed technique for repair of right superior vena cava draining to left atrium. Ann Thorac Surg 55: 1568-70.

3. Magalhães SP, Moreno N, Loureiro M, Franca M, et al. (2016) Anomalous pulmonary venous connection: an underestimated entity. Rev Port Cardiol 35: 637-718.

4. Craig JMB, Darling RC, Rothney WB (1957) Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 6: 44-64.

5. Kim TH, Kim YM, Suh CH, Cho DJ, Kim WH, et al. (2000) Helical CT angiography and three-dimensional reconstruction of total anomalous pulmonary venous connections in neonates and infants. AJR 175: 1381-6.

6. Valsangia ER, Levasseur S, McCrindle BW, MacDonald C, Smallhorn JF, et al. (2003) Contraste enhanced MR Angiography of pulmonary venous abnormalities in children. Pediatr Radiol 33: 92-98.

7. Avellana P, Segovia J, Lópe F, Gómez-Buen M, García-Cosío C, et al. (2012) Hypertensión pulmonary embarazo. Cardiocore 47: 154-60.

8. Bharati S, Lev M (1973) Congenital anomalies of the pulmonary veins. Cardiovasc Clin 5: 23-41.

9. Reddy KP, Nagarajan R, Rani U (2015) Total anomalous pulmonary venous connection beyond infancy. Asian Cardiovasc Thorac Ann 19: 249-52.

10. Jian XH, Huang J, Ding Y (2012) Surgical Outcome of Isolated Total Anomalous Pulmonary Venous Connection in Adults: A 14-Year Experience. J Card Surg 27: 736-9.

11. Atik FA, Jaramillo M, Afiune JY, Caneo LF (2009) Complete repair of total anomalous pulmonary venous connection in adult patient. Rev Bras Cir Cardiovasc 24: 81-3.

12. Galiè N, Humbert M, Vachiery JL, Gibbs S (2016) ESC / ERS 2015 guide on diagnosis and treatment of pulmonary hypertension (Guía ESC/ERS 2015 sobre diagnóstico y tratamiento de la hipertensión pulmonar). Rev Esp Cardiol 69: 177-259.