Mixed Variety of Total Anomalous Pulmonary Venous Return in an Asymptomatic Newborn

Variedade Mista de Retorno Venoso Pulmonar Anômalo Total em Recém-Nascido Assintomático

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Case Report

Total anomalous pulmonary venous return has an incidence of 7–9 per 100,000 live births, accounting for 0.7–1.5% of all congenital heart diseases. The most common type is the supracardiac, with the mixed form being the rarest. Moreover, it is one of the very few venous malformations known to cause cyanosis.[1]

The authors describe the case of a term newborn girl, whose first fetal scan was carried out only after the 20th gestational week. Fetal scans and laboratory work-up were normal and she was delivered vaginally at 38 weeks gestation. She was asymptomatic and remained at her mother's side. At 48 hours post-delivery, routine pulse oximetry was carried out, which showed peripheral oxygen saturation of 87%, with no signs of respiratory distress.

Transthoracic echocardiogram showed dilated right chambers, right-to-left shunt across a medium sized atrial septal defect, absence of normally draining pulmonary veins and posterior venous conduit draining to the right atrium (Figure 1). To better define the malformation, a coronary computed tomography angiography scan was carried out, which showed a

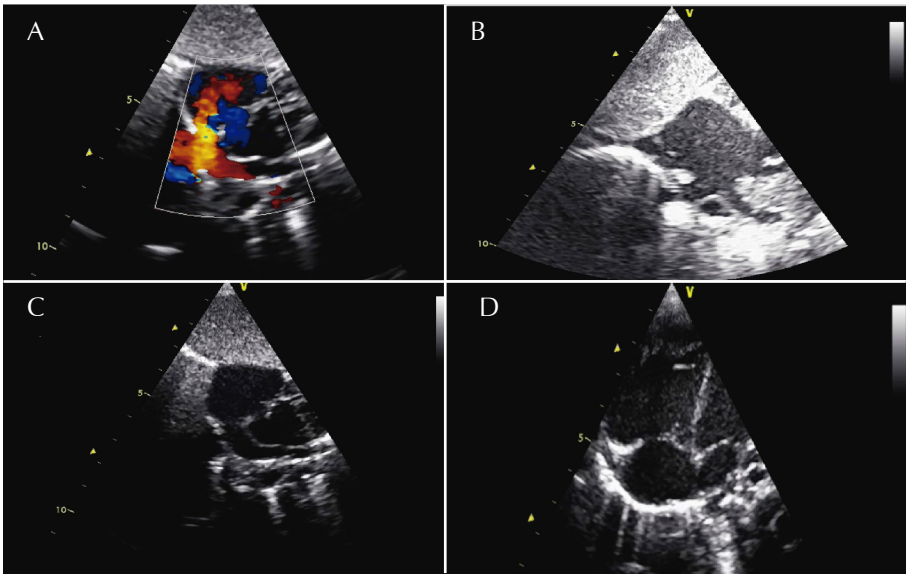

Figure 1 – *A) Subcostal view of transthoracic echocardiography with color Doppler revealing right-to-left shunt in the atrial septal defect (blue) and an anomalous pathway from a retro-atrial conduit emerging into the right atrium (red); B) Subcostal "bicaval" view with a right merging vase in the superior vena cava just before entering the right atrium; C) Anomalous retro-atrial conduit emerging into the right atrium; D) Apical four chamber view with dilated right sided chambers.*

Keywords

Congenital Heart Disease, Cyanotic Heart Disease; Paediatric Cardiology; Total Anomalous Pulmonary Venous Return; Newborn.

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mixed type of total anomalous pulmonary venous return, where the left pulmonary veins and the right inferior vein drained into the right atrium via a common trunk and right superior and middle pulmonary veins drained to the superior vena cava (Figure 2). Surgical correction was carried out at the age of 3 months, confirming the coronary computed tomography scan findings and with an excellent corrective result.

Discussion

The mixed variant or type IV of Darling's Classification of total anomalous pulmonary venous return is the rarest form of this entity. The surgical mortality rate is higher in this form, as are other related factors such as younger age at presentation and the presence of obstructive pulmonary veins.[2] Other than the early clinical presentation, our patient had no other associated anomalies, with an overall good outcome.

Prenatal fetal echocardiography is challenging, whereas postnatal echocardiography has a 97% sensitivity and specificity.[3] In our case, the diagnosis was postnatally, later confirmed by computed tomography angiogram and surgical findings.

Pulse oximetry prior to maternity discharge is an helpful tool to screen for asymptomatic congenital cardiac conditions.[4]

Authors' contributions

Research creation and design: Cláudio Henriques, Patrícia Silva, Andreia Palma. Data acquisition: Cláudio Henriques, Patrícia Silva. Manuscript writing: Cláudio Henriques, Andreia Palma. Intellectual support and revision: António Pires, Helena Andrade.

Conflict of interest

The authors declare that there is no conflict of interest regarding this manuscript.

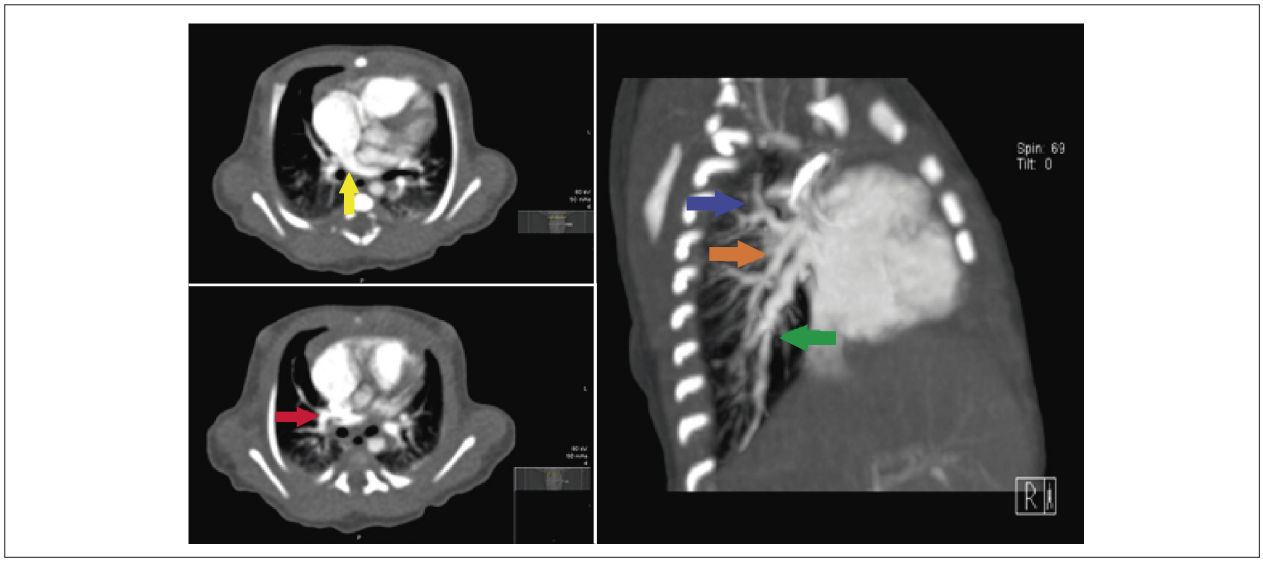

Figure 2 – *Axial view of the chest Computed Tomography Angiogram (left sided images) showing a common trunk coming from the left pulmonary veins and draining into the right atrium (yellow arrow) and the right inferior pulmonary vein merging into the aforementioned common trunk (red arrow). Right anterior longitudinal view of the chest Computed Tomography Angiogram (right image) revealing the right superior (blue arrow) and right middle (orange arrow) pulmonary veins flowing into the superior vena cava and the right inferior pulmonary vein merging into the common trunk, which in turn drains into the right atrium. The green arrow is highlighting the right inferior pulmonary vein.*

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