A unique congenital aorto-caval fistula: multi-modality imaging role for successful treatment

Miguel Nobre Menezes¹*, Ana G. Almeida², Ángelo Nobre⁴, and Luís Brás Rosário⁵,⁶

¹Cardiology Department, University Hospital Santa Maria, CHLN, Hospital de Santa Maria, Avenida Prof Egas Moniz, Lisbon 1649-035, Portugal; ²Faculty of Medicine, University of Lisbon, Lisbon, Portugal; ³Cardiology Department, University Hospital Santa Maria, CHLN, Lisbon 1649-035, Portugal; ⁴Cardiothoracic Surgery Department, University Hospital Santa Maria, CHLN, Lisbon 1649-035, Portugal; ⁵Cardiology Department, Santa Maria University Hospital, Lisbon, Portugal; and ⁶Faculty of Medicine/Cardiology Center University of Lisbon, University of Lisbon, Portugal

* Corresponding author. Tel: +351 217805351; Fax: +351 218860151. Email: mnmenezes.gm@gmail.com

A 45-year-old female presented with slowly progressing right heart failure. The cardiac auscultation revealed a continuous murmur heard all over the precordium, louder over the right sternal border. Transthoracic and transoesophageal echocardiograms were performed. An abnormal fistulous path with its origin in the right sinus of Valsalva was visible (Panel A). However, its distal end could not be identified. Additionally, all four heart chambers were dilated, with preserved systolic function of both ventricles and the determination of Qp/Qs was approximately 1. She had mild pulmonary hypertension (maximal velocity of minimal tricuspid regurgitation was 2.9 m/s). A CT angiogram was performed and revealed a fistula with an aneurysm morphology originating from the right aortic sinus of Valsalva and ending in the anterior wall of the left superior vena cava, just above the right atrium. The right coronary artery emerged from the proximal end of the fistula (Panels B and C). The patient underwent surgical intervention (Panel D) without extra-corporeal circulation and the fistula was closed, leaving the proximal end intact to ensure right coronary artery patency (Panel D). The patient post-operative evolution was uneventful and at 3-month follow-up, she had no symptoms nor signs of heart failure.

Congenital aorto-caval fistulas are exceedingly rare. Anatomical characteristics are variable and every fistula is unique, resulting in heterogeneous clinical presentation. This case illustrates combining cardiac imaging techniques can deliver a precise anatomical and functional evaluation sufficient for diagnosis and therapeutic decisions. Thus, even in very rare cases, successful treatment can be achieved despite limited experience.