Persistent left superior vena cava draining into left atrium, unroofed coronary sinus, and coronary sinus atrial septal defect as rare cause for severe right heart dilatation: diagnostic steps and therapy

Uwe Speiser†*, Silvio Quick†, Veit Sandfort, Ruth H. Strasser, and Steffen Kolschmann

Department of Internal Medicine and Cardiology, Dresden University of Technology, Heart Center, University Hospital, Fetscherstraße 76, Dresden 01307, Germany

† Both authors contributed equally.

* Corresponding author. Tel: +49 351 450 25 248; fax: +49 351 4501702, Email: uwe.speiser@mailbox.tu-dresden.de

A 44-year-old female patient with dyspnoea New York Heart Association III was referred for diagnostics. Transthoracic echocardiography revealed dilated right ventricle (RV) and impaired right ventricular function with mild pulmonary hypertension. Subsequent contrast application from the right antecubital vein showed early contrast in right atrium (RA, Panel A and Supplementary data online, Movie S1) and moderate interatrial shunt. By contrast injection from the left antecubital vein, immediate contrasting of left atrium (LA) and left ventricle could be demonstrated with subsequent contrasting of RA and RV via atrial septal defect (ASD, Panel B and Supplementary data online, Movie S2). This suggested the existence of a persistent left superior vena cava (PLSVC) draining into the LA. Coronary artery disease was invasively excluded. Right heart catheterization demonstrated a left-right shunt (Qp/Qs ratio 2.2). A catheter was pushed into the LA passing an ASD, and PLSVC joining into the LA was identified (Panel C, asterisk indicates PLSVC, and Supplementary data online, Movie S3). Transoesophageal echocardiography confirmed ASD located in the infero-posterior region of interatrial septum (Panel D, white arrow indicates ASD). Coronary sinus appeared unroofed. Cardiac magnetic resonance (CMR) also verified the large ASD and the unroofed coronary sinus. Cine sequences showed an interatrial crossed shunt (Panel E, arrow matches ASD, asterisk shows PLSVC). MR angiography confirmed echocardiographic suspicion (Panel F, asterisk indicates PLSVC). Eighteen month later, the patient agreed with surgical treatment. The ASD was closed by a patch (Panel G, arrow indicating ASD patch) and PLSVC placed into the RA (Panel H, asterisk indicates PLSVC). Further clinical course proceeded regular. This case demonstrates the pivotal role of multimodal imaging in congenital heart disease to make accurate diagnosis and initiate sufficient therapy.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.