Use of multi-modality imaging in a patient with a persistent left superior vena cava, partial anomalous pulmonary venous connection, and sinus venosus-type atrial septal defect

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A 45-year-old woman with a history of congenital heart disease that was not previously well defined presented with progressive dyspnoea on exertion for 1 year. Three-dimensional (3D) transthoracic (TTE) and transoesophageal (TEE) echocardiography with agitated saline injection (bubble study) were performed. A severely dilated coronary sinus (CS) was visualized measuring 3.5 cm in its largest diameter with a present CS roof separating the CS from the left atrium (Figure A). Bubbles from the left upper extremity were evident filling first the CS then the severely dilated right atrium and right ventricle, respectively (Figure B), suggesting a persistent left superior vena cava (SVC). The CS, left upper and lower pulmonary veins and right lower pulmonary vein were identified draining into the left atrium (Figure C). Despite multiple attempts, the right upper pulmonary vein (RUPV) and the SVC-right atrial junction could not be visualized on TTE/TEE. Subsequent computed tomography with 3D/4D reconstruction confirmed the persistent left SVC without a bridging innominate vein (Figure D), emptying into the CS (Figure E). The RUPV formed a vascular confluence with the right SVC (Figure D) joining superiorly the right atrium creating a partial anomalous pulmonary venous connection and large sinus venosus-type atrial septal defect (ASD) (Figure F). Surgical correction was performed by construction of an intracardiac baffle redirecting the RUPV into the left atrium and patch closure of the ASD. Our case identifies the successful use of multi-modality imaging to identify and understand complex congenital anatomy.

ASD, atrial septal defect; CS, coronary sinus; CSR, coronary sinus roof; LA, left atrium; LLPV, left lower pulmonary vein; L SVC, left superior vena cava; LUPV, left upper pulmonary vein; LV, left ventricle; RA, right atrium; RLPV, right lower pulmonary vein; R SVC, right superior vena cava; RUPV, right upper pulmonary vein; RV, right ventricle.

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