Unanticipated stenosis of the systemic arterial supply in scimitar syndrome

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A 28-year-old woman, with known Scimitar syndrome, was evaluated for moderate exercise intolerance. Examination was notable for grade 2/6 systolic murmur over the pulmonary area. The electrocardiogram was unremarkable. Chest X-ray demonstrated dextroposition of the heart with a shadow suggestive of the scimitar vein. Transthoracic echocardiogram was technically challenging with no evidence of pulmonary hypertension or intracardiac shunts. Cardiovascular magnetic resonance showed moderately dilated right ventricle with normal systolic function and hypoplasia of the right lung and right pulmonary artery (PA) (Supplementary material online, Video 1). The right pulmonary veins (PVs) joined in a confluence and drained into the right atrium (RA) flush with the inferior vena cava junction above the diaphragm. The left PVs drained normally into the left atrium. The Qp/Qs ratio was 1.5:1. (Panels A–C, Supplementary material online, Video 2). The entire right lung was supplied by the right PA except for a small area of the basal zone which was fed by small systemic arterial collaterals arising from the celiac axis. These collaterals demonstrated multiple severe stenoses and post-stenotic aneurysmal dilatation throughout their course. This was confirmed by cardiac computed tomography (CCT) (Panels D and E). CCT revealed normal bronchial anatomy and lung parenchyma.

A decision was made to proceed with surgical correction. The RA was incised to create an atrial septal defect followed by redirecting the right PVs confluence to the defect. Basal lung resection was deferred as the lung parenchyma was normal. Ligation of anomalous collaterals was not performed given concerns of causing pulmonary infarction. Recovery was uneventful.

(A–C) CMR showing right PVs confluence (arrows) draining into the RA with left PVs draining normally into the left atrium. Maximum intensity projection (D) and three-dimensional reconstruction CCT (E) showing stenotic anomalous systemic arterial collaterals (arrows) with aneurysmal dilatation. IVC, inferior vena cava; LA, left atrium; LUPV/LLPV, left upper/left lower pulmonary vein; RV, right ventricle.

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

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