A 6-year-old girl presented with progressive dyspnoea and cyanosis. Examination revealed grade 1 clubbing, cyanosis (saturation of 85%), right ventricular (RV) heave, and loud S2. ECG and chest radiograph showed evidence of RV pressure overload (Supplementary material online, Figures S1 and S2). Transthoracic echocardiography demonstrated a membrane dividing the left atrium (LA) into a proximal accessory chamber (AC) and a distal main chamber, consistent with classic cor triatriatum (Panel A—parasternal long axis view, Supplementary material online, Videos S1 and S2). All four pulmonary veins drained into the AC. The membrane showed a restrictive aperture with mean pressure gradient of 13 mmHg (Panel B—colour flow imaging in apical 4-chamber view; Panel C—continuous wave Doppler). The inter-atrial septum between the AC and right atrium (RA) was intact; however, it had a large defect with left to right shunting at the level of the LA main chamber. The RA, RV, and pulmonary arteries were grossly dilated, indicating severe pulmonary hypertension. A persistent left superior vena cava (PLSVC) was noted. For evaluation of cyanosis, contrast echocardiography was performed by injecting agitated saline into the left antecubital vein. This revealed sequential opacification of coronary sinus (CS), LA main chamber, LV and RA, indicating a right to left shunt across an unroofed CS (Panel D, Supplementary material online, Video S3). ECG-gated, contrast-enhanced, 64-slice multidetector CT confirmed presence of a PLSVC (blue stars in Panel E) draining into a dilated, completely unroofed CS (Kirklin type I) (red stars in Panel E; Panel F—volume rendered image) and a 2-mm-thick membrane with a 7-mm fenestration near a huge LA appendage (Panel G).

This case illustrates a unique physiology of coexistent opposing shunts at the level of the LA. Association of classic cor triatriatum with type I unroofed CS is rare; recognition of the unroofing may be tricky and requires high index of suspicion along with use of multimodality imaging. Diagnosis is important due to attendant risk of cyanosis, systemic emboli and brain abscess. Early surgical repair with excision of the membrane and intra-atrial rerouting of PLSVC to RA with a pericardial patch has shown excellent outcomes.

Supplementary material is available at European Heart Journal online.

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