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CARDIOVASCULAR FLASHLIGHT
Congenital heart disease

Tricuspid atresia in adulthood

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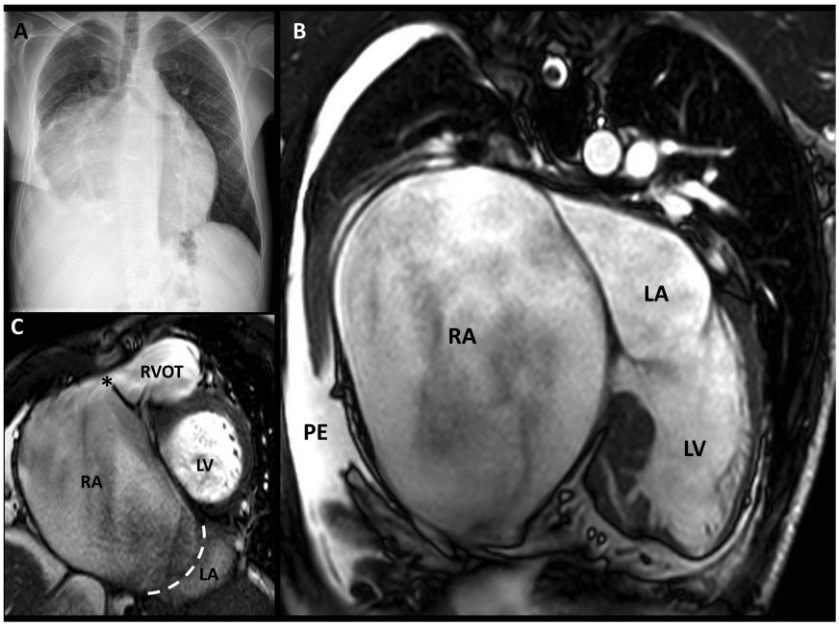
A 40-year-old man presented to the emergency department with worsening exertional breathlessness, orthopnoea, and leg swelling over the last 2 weeks. Examination revealed large ascites and hydrocele. He was known to have tricuspid atresia type IB and ostium secundum atrial septal defect (ASD) treated surgically with a Waterston shunt (an anastomosis between the ascending aorta and the right pulmonary artery) at 3 months of age, followed by Fontan–Björk procedure—connecting right atrial appendage and right ventricular infundibulum—and ASD closure at 5 years of age. The patient was regularly followed-up at a specialized grown-up congenital heart disease service but repeatedly refused total cavopulmonary connection. During the last 5 years was recurrently admitted to the hospital due to refractory right-sided heart failure. He was on rivaroxaban 20 mg once daily (primary prevention of stroke and thromboembolic events), metoprolol 50 mg twice daily (rate control of permanent atrial fibrillation), and furosemide 125 mg twice daily (maximally tolerated dose), with persistent symptoms of congestion at rest or with activities of daily living (INTERMACS profile 4).

Electrocardiography showed atrial fibrillation (Supplementary material online, *Figure S1*). Chest radiography revealed severe



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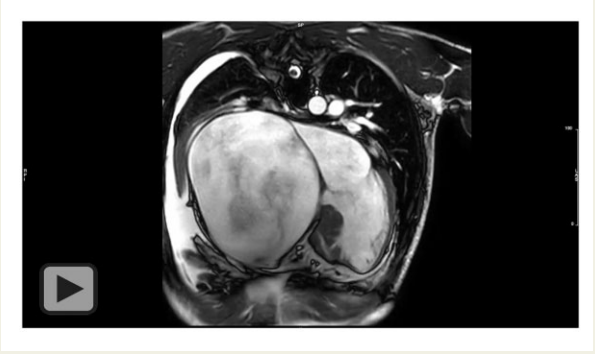

Video 1 Cardiovascular magnetic resonance cine balanced steady-state free precession, horizontal long-axis view.

cardiomegaly (*Panel A*). Cardiovascular magnetic resonance showed a giant right atrium (right atrial volume 1665 mL, normal <169 mL) with sluggish blood flow (*Panel B*; *Video 1*) and a patent Fontan–Björk connection (*Panel C*, asterisk). Left ventricular systolic function was low-normal and no other intracardiac shunts were identified.

Due to end-stage Fontan–Björk physiology causing refractory heart failure and portal hypertension, the patient is now awaiting orthotopic heart transplantation. Despite the growing number of people living with Fontan circulation in the adulthood, long-term mortality remains substantial and exposes these patients to a variety of complications. Namely, heart failure is among the leading causes of hospital admission in the Fontan population, and once apparent, it is harbinger of poor outcomes.

(*Panel A*) chest radiography showing massive cardiomegaly and a prominent right heart border that reflects enlargement of the right atrium. (*Panel B*) CMR cine b-SSFP image (horizontal long-axis view) showing a giant right atrium with sluggish blood flow, normal-sized left atrium, non-dilated left ventricle with excessive trabeculation, and pleural effusion. *Panel C*: CMR b-SSFP image (short-axis view) showing patent Fontan–Björk connection connecting right atrial appendage and right ventricular infundibulum. bSSFP, balanced steady-state free precession; CMR, cardiovascular magnetic resonance; LA, left atrium; LV, left ventricle; PE, pleural effusion; RA, right atrium; RVOT, right ventricular outflow tract.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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