Tricuspid Atresia in Adulthood

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Short abstract: Patients with tricuspid atresia can reach their adulthood after Fontan operation. The major challenge is to maintain quality of life for the failing Fontan adult survivors, as well as extending their life expectancy.

A 40-year-old man presented to the emergency department with worsening exertional breathlessness, orthopnea and leg swelling over the last two 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 specialised grown-up congenital heart disease (GUCH) 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 (Online Figure S1). Chest radiography revealed severe cardiomegaly (Panel A). Cardiovascular magnetic resonance showed a giant right atrium (right atrial volume 1,665 mls, normal <169 mls) with sluggish blood flow (Panel B; supplementary online material, Video S1) 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 outcome.

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**Figure legend**

**Figure 1.** 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, nondilated 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 online material**

Video S1. CMR cine bSSFP, horizontal long-axis view.

Figure S1. 12-lead ECG showing atrial fibrillation.

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