

Tricuspid Atresia in Adulthood

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

27

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

41 Electrocardiography showed atrial fibrillation (Online Figure S1). Chest radiography revealed severe
42 cardiomegaly (Panel A). Cardiovascular magnetic resonance showed a giant right atrium (right atrial
43 volume 1,665mls, normal <169mls) with sluggish blood flow (Panel B; supplementary online
44 material, Video S1) and a patent Fontan-Björk connection (Panel C, asterisk). Left ventricular
45 systolic function was low-normal and no other intracardiac shunts were identified.

46 Due to end-stage Fontan-Björk physiology causing refractory heart failure and portal hypertension,
47 the patient is now awaiting orthotopic heart transplantation. Despite the growing number of people
48 living with Fontan circulation in the adulthood, long-term mortality remains substantial and exposes

49 these patients to a variety of complications. Namely, heart failure is among the leading causes of
50 hospital admission in the Fontan population, and once apparent, it is harbinger of poor outcome.

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53 **Consent:** The patient has given his consent for the use of his medical data and images.

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55 **Figure legend**

56 **Figure 1.** *Panel A:* chest radiography showing massive cardiomegaly and a prominent right heart
57 border that reflects enlargement of the right atrium. *Panel B:* CMR cine b-SSFP image (horizontal
58 long-axis view) showing a giant right atrium with sluggish blood flow, normal sized left atrium,
59 nondilated left ventricle with excessive trabeculation, and pleural effusion. *Panel C:* CMR b-SSFP
60 image (short axis view) showing patent Fontan-Björk connection connecting right atrial appendage
61 and right ventricular infundibulum.

62 bSSFP, balanced steady-state free precession; CMR, cardiovascular magnetic resonance; LA, left
63 atrium; LV, left ventricle; PE, pleural effusion; RA, right atrium; RVOT, right ventricular outflow
64 tract.

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66 **Supplementary online material**

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

68 Figure S1. 12-lead ECG showing atrial fibrillation.

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70 The authors do hereby declare that all illustrations and figures in the manuscript are entirely
71 original and do not require reprint permission.

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