A 48-year-old woman presented with progressive exercise intolerance and paroxysmal atrial fibrillation. The past history was remarkable for severe Ebstein’s anomaly and hypoplastic right ventricle, for which she had undergone placement of a 30 mm Carpentier-Edwards right atrial to right ventricular outflow tract conduit at 20 years of age. She subsequently required conduit revision with a 26 mm pulmonary cryopreserved homograft due to symptomatic conduit obstruction at the age of 24 years. Physical examination revealed mid-systolic click, widely split S2 and Grade 2/6 ejection systolic murmur at the heart base. Transthoracic echocardiography was technically difficult. Transoesophageal echocardiography demonstrated the conduit arising from the mid-portion of right atrial free wall with a moderately regurgitant bioprosthetic valve measuring 15 mm proximally. The conduit progressively narrowed distally to 7 mm just before the right ventricular outflow tract anastomosis. Ebstein’s malformation with moderate tricuspid valve regurgitation was noted (Figure 1A; see Supplementary data online, Video S1). Cardiovascular magnetic resonance demonstrated findings similar to echocardiography. Additionally, severe right atrial dilation and a small functional right ventricle with mildly reduced systolic function (ejection fraction = 43%) were seen (Figure 1B and C; see Supplementary data online, Videos S2–S4). Balloon occlusion of the conduit for up to 5 min at cardiac catheterization had no impact on the patient’s haemodynamics, suggesting that the conduit was not contributing importantly to right atrial–right ventricular flow. A decision was made to proceed with conduit excision and tricuspid valve repair.

In symptomatic patients with severe tricuspid valve malformation, a modified Fontan procedure offers a promising palliation avoiding surgery in early childhood.

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

Figure 1 (A) Transoesophageal echocardiography modified apical four-chamber view showing the proximal conduit with a bioprosthetic valve (red arrowhead). The conduit progressively narrows distally to 7 mm before its right ventricular outflow tract anastomosis (yellow arrow). RA, right atrium; RV, right ventricle. (B) Cardiovascular magnetic resonance modified four-chamber view showing the proximal (black arrow) and distal (yellow arrow) conduit anastomosis (see text for details). LV, left ventricle; RA, right atrium; RV, right ventricle. (C) Cardiovascular magnetic resonance three-dimensional reconstruction model showing the conduit anastomosis between the right atrium and right ventricular outflow tract. RA, right atrium; RVOT, right ventricular outflow tract.