Successful reoperation of the valveless calcified right atrium to right ventricle conduit in an adult patient with tricuspid atresia after Fontan procedure

Justyna Rybiccka*, Mirosław Kowalski, Jacek Różański and Piotr Hoffman

Abstract

A 31-year old woman with tricuspid atresia, after Bjork Fontan modification and reoperation due to homograft narrowing, was evaluated because of progressive exercise intolerance; she was planning a pregnancy. The right atrium (RA) to right ventricle (RV) homograft narrowing and calcification with significant reversal flow were found. Successful reoperation was performed—calcified conduit was excised and extracardiac pulmonary homograft tipped with vascular prosthesis was implanted between the RA and the RV.

Keywords: Tricuspid atresia • Fontan operation • Right atrium • Right ventricle • Homograft

INTRODUCTION

Narrowing either of the systemic venous connection or, as in the case of the Bjork modification Fontan procedure, the right atrium (RA) to right ventricle (RV) conduit is an important cause of the failing Fontan [1]. The possible management options in this case include: percutaneous or surgical removal of the obstruction (e.g. valve implantation) and conduit replacement or conversion to a total cavopulmonary connection (TCPC) [1–4]. We postulate that it is not always safe to put valve percutaneously into conduit and, at the same time, that reoperation gives excellent results.

CASE REPORT

The 31-year old female patient with tricuspid atresia underwent at the age of 6 Fontan in Bjork modification surgery with the use of aortic homograft. At the age of 14, she underwent reoperation because of homograft narrowing. The excision of the calcified valve causing stenosis with subsequent connection of the remaining part of the homograft with pericardial tunnel was done. Recently, she reported progressive exercise intolerance (NYHA Class II/III). She was planning a pregnancy. Echocardiography discovered the conduit diameter of 5–7 mm with peak and a mean gradient of 12 and 7 mmHg. There was also a diastolic reversal flow through the conduit to the dilated RA (69 × 51 mm) and widened inferior vena cava (36 mm) with a decreased respiratory variation. The cardiopulmonary exercise test confirmed the patient’s poor exercise capacity (peak workload 2.98 metabolic equivalents, peak VO2 10.44 ml/kg/min). In cardiac magnetic resonance imaging, small left ventricle with preserved systolic function and small RV [end-diastolic volume (EDV) 62 ml, end-systolic volume 35 ml, EDV/body surface area 41 ml/m²] with mildly impaired systolic function (systolic volume 26 ml, ejection fraction 43%) were present. It also showed largely dilated RA and valveless conduit connecting RA to RV with a minimal diameter of 19 × 13 mm (Fig. 1). The reversal flow in the conduit had a volume of 16 ml, which constituted 26% of RV EDV. Catheterization showed the mean RA pressure of 20 mmHg and RA–RV end-diastolic gradient of 14 mmHg. Both surgical treatment and percutaneous valve implantation were considered. Before a final decision concerning treatment was made, computed tomography had been performed. It revealed severe calcification of the conduit (Fig. 2). The idea of percutaneous valve implantation was abandoned due to high risk of calcified pericardial tunnel perforation. The patient was qualified for surgical treatment. Severely calcified RA–RV conduit adhering to the sternum was excised. Extracardiac pulmonary homograft 25 mm was implanted to the RA and subsequently connected to the RV with the use of vascular prosthesis 24 mm. In the post-surgical 1-month follow-up, she got improved. Her physical capacity substantially improved corresponding with high risk of calcified pericardial tunnel perforation. The patient was qualified for surgical treatment. Severely calcified RA–RV conduit adhering to the sternum was excised. Extracardiac pulmonary homograft 25 mm was implanted to the RA and subsequently connected to the RV with the use of vascular prosthesis 24 mm. In the post-surgical 1-month follow-up, she got improved. Her physical capacity substantially improved corresponding with high risk of calcified pericardial tunnel perforation. The patient was qualified for surgical treatment. Severely calcified RA–RV conduit adhering to the sternum was excised. Extracardiac pulmonary homograft 25 mm was implanted to the RA and subsequently connected to the RV with the use of vascular prosthesis 24 mm. In the post-surgical 1-month follow-up, she got improved. Her physical capacity substantially improved corresponding with high risk of calcified pericardial tunnel perforation.

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DISCUSSION

In the case presented here, 25 years after conduit implantation, no Fontan circulation complications were found. Thus, when sufficiently developed RV is present, the RA–RV conduit may be considered as a reasonable choice [5, 6]. Frequent conduit narrowing is presumably due to the use of an aortic homograft and pericardial conduit that are prone to calcification (secondary to a high amount of elastic fibres and total tissue calcium) [7, 8]. Considerably, better results are obtained in the case of a pulmonary homograft implantation. What should be born in mind is that the risk of conduit disruption during sternotomy is high. Therefore, before sternotomy is performed, it is recommended to establish cardiopulmonary bypass via femoral vessels. An alternative method for removing homograft narrowing is percutaneous valve implantation into the narrowed conduit [3]. In our case, we did not choose to go for interventional treatment because of severe calcifications of the homograft and a high risk of conduit perforation. Instead, we chose reoperation—we abandoned TCPC conversion and decided to replace the RA–RV conduit because of sufficiently developed RV with a reasonably good contractility. It allowed preservation of biventricular circulation and came out very well. Thus, when feasible, what we advocate here is qualification of patients with conduit narrowing after Fontan operation for surgery treatment. In the case of Bjork modification if only RV size and function are adequate, we recommend to replace the RA–RV conduit rather than to perform TCPC conversion.

ACKNOWLEDGEMENTS

The authors thank Dr Ilona Michalowska for preparing Figure 2 and Dr Łukasz Mazurkiewicz for preparing Figure 1.

Conflict of interest: none declared.

REFERENCES


