CASE REPORT - CONGENITAL

Successful Bentall procedure in a patient with a Fontan circulation

Veronica Spadottoab, Hideki Uemurac,* and Anselm Uebinga

a Heart Division, Royal Brompton Hospital, London, UK
b Department of Thoracic and Cardiovascular Sciences, University of Padua, Padua, Italy
c Congenital Heart Disease Center, Nara Medical University, Nara, Japan

Abstract

A 33-year old male patient with absent left atrioventricular connection and double outlet left ventricle developed severe aortic root dilatation and aortic regurgitation. He had undergone initial banding of the pulmonary arterial trunk and atrial septectomy, eventually followed by the bidirectional Glenn procedure. At the time of the total cavopulmonary connection (at 27 years of age), his ascending aorta was plicated and wrapped. Subsequently, the aortic root below the wrapped ascending aorta showed further dilatation. The Bentall procedure was successfully carried out.

Keywords: Fontan • Bentall • Aortic root dilatation

INTRODUCTION

Several revisional surgical procedures have been described in patients with a Fontan circulation for such problems as arrhythmias, venous pathway obstruction and atrioventricular valve regurgitation. Aortic root dilatation requiring the Bentall procedure subsequent to total cavopulmonary connection (TCPC) has remained a rare situation [1]. Thus, we describe a case herein.

CASE REPORT

A male patient with absent left atrioventricular connection and double outlet left ventricle (dominant morphological left ventricle with a rudimentary morphological right ventricle on the left side) initially underwent banding of the pulmonary arterial trunk and atrial septectomy in 1979 (at 18 months of age). The second operation had not been carried out until 1990, when the bidirectional Glenn procedure was employed at the age of 11 years. An additional forward flow had been maintained across the pulmonary arterial banding. He was referred to our clinic, at 23 years of age, to assess his suitability for Fontan circulation. At that time, his peripheral oxygen saturation was 86%, and atrial pressure was 6 mmHg with a trans pulmonary pressure gradient of 4 mmHg. On cardiac magnetic resonance (CMR), left ventricular end-diastolic volume index (LVEDVI) was 200 ml/m² and left ventricular ejection fraction (LVEF) 42% with globally impaired contraction. The aortic root sinus was 43 mm and the ascending aorta 57 mm in diameter with mild plus AR. LVEDVI remained at 196 ml/m² with an LVEF of 26%. Reoperation was not immediately carried out because of poor ventricular contractility. Six years after TCPC completion, AR progressed to a significant degree, with LVEDVI 177 ml/m², LVEF 38% and Valsalva sinus up to 67 mm in diameter (Fig. 1). Regurgitant fraction was 38% on CMR. At 33 years of age, the Bentall procedure took place uneventfully using a composite graft bearing a 31-mm St Jude valve. All the aortic valvular leaflets had become considerably degenerative, so that sparing his native valve did not appear to be a durable long-term solution. After coming off cardiopulmonary bypass, central venous pressure was 8–10 mmHg. The only technical issue encountered was reconstruction of the coronary buttons, because they needed afferent mobilization of 18 mm or greater because of differences in diameter between the original (the dilated ascending aortic root) and the reconstructed (the tube graft) wall. The coronary artery arising from the left lateral aspect (morphologically Sinus 1) was small compared with the other coronary artery, and coursing just in front of the ventriculoarterial junction. Given the anatomical feature, mobilization of the coronary button was limited. Fortunately, reconstruction of the coronary arteries turned out to be efficient without use of an inter-position graft (Fig. 2).

The postoperative course was smooth with a duration of stay in the intensive care unit of 24 h. Eight months later, the patient was doing well with an international normalization ratio of 3–3.7 on warfarin.

DISCUSSION

Primary intrinsic aortopathy with progressive aortic dilatation represents a new emerging concept in adult patients with congenital
heart disease, apart from Marfan syndrome, Turner syndrome, bicuspid aortic valve, coarctation of the aorta or hypoplastic left heart syndrome [2]. To the best of our knowledge, only a few case reports have been published with regard to aortic or neoaortic root surgery after Fontan completion [1, 3, 4]. The latter two reports [3, 4] were mainly for the neoaortic (anatomically pulmonary) root after the Norwood procedure. Only Egan et al. [1] described the combination of aortic root replacement using a prosthesis and the Fontan circulation.

It remains debatable whether the general guideline for replacement of ascending aortic aneurysm should be applied also in congenital heart disease; it is known that the ascending aorta is greater than normal from the beginning in some congenital situations, such as tetralogy of Fallot. Therefore, surgical indication should include not only the exact figure of the dilated ascending aorta, but also other complementary factors (increase in size of the structure or AR at sequential observation, etc.). In our patient, there was no reservation regarding surgical indication: its quite large geometry, its form, minor interval change in size as well as presence of AR. The mechanism of the progressive severe dilatation of the aortic root was most likely the reduced compliance of the ascending aortic wall subsequent to plication and wrapping. In this respect, the all part of the ascending aorta, including the aortic root, should have better been replaced. Nonetheless, doing both the Bentall procedure and completion of Fontan circulation at the same time could have led to severe ventricular impairment after extensive surgery. In reality, his ventricle struggled more or less after offloading by Fontan completion, even being carried out in the form of a less invasive method. It was readily considered that long-standing cyanosis and volume load through the additional forward flow were the causes of significant impairment of ventricular function. We decided to wait until ventricular performance recovered after offloading, because improvement could take longer than a year [5]. Fortunately, his dilated aortic root did not experience rupture or dissection [1], before the Bentall procedure. Retrospectively, the better approach in this particular patient could have been ascending aortic and aortic root replacement with the aortic valve spared, together with closure of the pulmonary arterial trunk as the first-stage procedure, followed by completion of the TCPC as the second stage.

CONCLUSION

The Bentall procedure was performed safely in an adult patient with a Fontan circulation. Strategy for treating a dilated aortic pathway in a univentricular physiology requires careful attention and thoughtful consideration.

Conflict of interest: none declared.

REFERENCES


