Aortopathy in an Adult With Tricuspid Atresia and Left Ventricular Non-Compaction After Fontan Procedure

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Although left ventricular non-compaction (LVNC) was originally described as an isolated cardiomyopathy, LVNC has been observed in association with several congenital heart diseases (CHD).1 Reports of an association with tricuspid atresia (TA), however, and of successful Fontan procedure for LVNC and TA, are scarce.2 In contrast, a new concept of “aortopathy” in which aortic dilation, aneurysms, dissections, and/or aortic

**Figure 1.** (A) Three dimensional image in the short axis view at the level of the aortic valve, showing a space between the three cusps located centrally (asterisk) during the end-diastolic phase because of a dilated aortic root. (B) Color Doppler image in the short axis view at the level of the aortic valve, showing aortic regurgitant jet from the central space. (C) Color Doppler image in the five chamber view, showing moderate-severe aortic regurgitation and the presence of a color (white arrow) in the thick LV wall intertrabecular recesses. (D) Color Doppler image in the long axis view, showing moderate-severe aortic regurgitation. Ao, aorta; AR, aortic regurgitation; LV, left ventricle.
regurgitation (AR) occur, resulting in a negative impact on systemic ventricular function, has been recognized in some CHD including coarctation of the aorta, bicuspid aortic valve, and conotruncal abnormalities such as tetralogy of Fallot. Increasing evidence has suggested that functional single ventricle after Fontan procedure should be categorized as an aortopathy. Herein we report the case of an adult after Fontan procedure for TA and LVNC in whom surgical intervention was necessary due to a progressively dilated aorta with AR.

A 36-year-old man was admitted to hospital for management of heart failure. He was diagnosed with TA, pulmonary valve absence, ventricular septal defect, atrial septal defect, single coronary artery, coronary arteriovenous fistula and LVNC. Extra-cardiac total cavopulmonary connection (TCPC) using a 22-mm Gore-Tex conduit with 4.5-mm fenestration had been performed when he was 27 years old, preceded by a bi-directional Glenn procedure at the age of 24 years. Cardiac catheterization performed at the age of 28 years indicated a central venous pressure of 8 mmHg without any pulmonary artery conduit or suprarenal vena cava (Fontan route) stenosis. Aortic saturation was 92% and cardiac index was 3.2 L/min/m². Angiography showed that the size of the aortic valve and sinus of Valsalva had increased, but that the descending aorta had not changed compared with that 9 years earlier (aortic valve, from 19 mm to 23 mm; sinus of Valsalva, from 42 mm to 64 mm; descending aorta, from 21 mm to 22 mm). Sellers’ grade III AR was observed. LV end-diastolic volume was 98 mL/m² and LVEF was reduced at 32%. The patient underwent aortic valve replacement and aortoplasty, reducing the size of the ascending aorta.

Dilatation of the aorta and/or AR are rarely encountered in patients with functional single ventricle after Fontan procedure. A few cases of dissection of the aorta or progressive aortic dilation with valvular regurgitation have been published. When volume loading and hypoxia persist for a considerable period, and the time since Fontan procedure is long, or when the aortic valve is bicuspid, the aortic root may become dilated and result in aneurysmal changes. Recent reports have shown that the elasticity of the ascending aorta may decrease and the wall stiffness increase in Fontan patients. Morphologic abnormalities of the ascending aortic wall were demonstrated in single ventricle and TA. More recently, older age at Fontan, male sex, elevated blood pressure, and LV morphology have been shown to be associated with severe aortic dilation. Aortic dilation with valve dysfunction is one of the long-term problems after Fontan procedure and this should be evaluated during follow-up, especially in adult male patients.

Disclosures
The authors declare no conflict of interest.

References