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

Tomotaka Murakami, MD; Yoshiki Mori, MD, PhD; Nao Inoue, MD, PhD; Sachie Kaneko, MD; Yasumi Nakashima, MD

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).\textsuperscript{1} Reports of an association with tricuspid atresia (TA), however, and of successful Fontan procedure for LVNC and TA, are scarce.\textsuperscript{2} In contrast, a new concept of “aortopathy” in which aortic dilation, aneurysms, dissections, and/or aortic

![Figure 1](https://example.com/figure1.png)

**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 absences, 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 superior vena cava (Fontan route) stenosis. Aortic saturation was 92% and cardiac index was 3.2 L/min/m². Angiography showed no AR. The postoperative clinical course had been good with warfarin and diuretics. He had New York Heart Association functional class IA, and worked full-time with edema. Despite increasing the oral dose of diuretics, the edema did not improve and he was admitted to hospital.

Echocardiography showed moderate-severe AR without coaptation of the cusps, and a reduced left ventricular ejection fraction (LVEF) of 31% (Figure 1). Contrast computed tomography (CT) showed that the spongy left ventricular (LV) wall was very thick and the sinus of Valsalva was prominently dilated (Figure 2). Serum B-type natriuretic peptide and atrial natriuretic peptide were increased at 1,800 pg/mL and 1,230 pg/mL, respectively.

After treatment with milrinone and i.v. furosemide drip, the edema improved and he lost 4 kg over 2 weeks compared with the admission weight. Cardiac catheterization was then performed. Central venous pressure was 16 mmHg and LV end-diastolic pressure was elevated at 15 mmHg. Aortic saturation was 92% and cardiac index was reduced to 1.8 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

1. Stahli BE, Gebhard C, Biagggi P, Klaassen S, Valsangiocomo Buechel E, Attenhofer Jost CH, et al. Left ventricular non-compaction: Prevalence in congenital heart disease. Int J Cardiol 2013; 167: 2477–2481.
2. Nguyen HH, Khan R, Silverman NH, Singh GK. Tricuspid atresia with non-compaction: An early experience with implications for surgical palliation. Pediatr Cardiol 2017; 38: 495–505.
3. Francois K. Aortopathy associated with congenital heart disease: A current literature review. Ann Pediatr Cardiol 2015; 8: 25–36.
4. Egan M, Phillips A, Cook SC. Aortic dissection in the adult Fontan with aortic root enlargement. Pediatr Cardiol 2009; 30: 562–563.
5. Erez E, Tam VK, Galliani C, Nancy AL, Peretti J. Valve-sparing aortic root replacement for patient with a Fontan circulation. J Heart Valve Dis 2012; 21: 175–180.
6. Myers KA, Leung MT, Potts MT, Potts JE, Sandor GG. Noninvasive assessment of vascular function and hydraulic power and efficacy in pediatric Fontan patients. J Am Soc Echocardiogr 2013; 26: 1221–1227.
7. Kojina T, Kuwata S, Kurishima C, Iwamoto Y, Saiki H, Ishido H, et al. Aortic root dilation and aortic stiffness in patients with single ventricular circulation. Circ J 2014; 78: 2507–2511.
8. Nowa K, Perloff JK, Bhuta SM, Laks H, Drinkwater DC, Child JS, et al. Structural abnormalities of great arterial walls in congenital heart disease: Light and electron microscopic analyses. Circulation 2001; 103: 393–400.
9. Kim YY, Rathod RH, Gauvreau K, Keenan EM, del Nido P, Geva T. Factors associated with severe aortic dilation in patients with Fontan palliation. Heart 2017; 103: 280–286.