Atrial fibrillation ablation without pulmonary vein isolation in an atriopulmonary Fontan circulation

John L. Fitzgerald, MBBS (Hons),* Nicholas J. Collins, MBBS,* James Leitch, MBBS,* Eugene Downar, MD,† Krishnakumar Nair, MD,† Nicholas Jackson, MBBS*

*From the *Department of Cardiology, John Hunter Hospital, Newcastle, Australia, and †Toronto General Hospital, University Health Network, Toronto, Canada.*

Introduction

Patients undergoing Fontan palliation for complex congenital heart disease (particularly those with earlier iterations of the procedure, such as the atriopulmonary [AP] Fontan) are prone to late arrhythmic events, most typically intra-atrial reentrant tachycardia (IART). The management of atrial fibrillation (AF) in the Fontan circulation is less well described. We present a case with a substrate-based approach to ablation of right-sided AF with organization of electrical activity to IART and subsequent successful ablation of this as well.

Case report

A 36-year-old woman presented to hospital with palpitations associated with lethargy, fatigue, and cyanosis. The past medical history was significant for double inlet left ventricle with ventricular septal defect, hypoplastic right ventricle, tricuspid atresia, and dextro-transposition of the great arteries. The patient was initially managed with pulmonary artery banding and a Blalock-Taussig shunt before undergoing Glenn shunt creation. The patient then proceeded to a modified AP Fontan shunt with Damus-Kaye-Stansel connection aged 7 years. Electrophysiology study and ablation had been successfully performed 6 years previously (at age 30) for IART (a peritricuspid annular circuit), with sotalol not tolerated.

Presently, the patient described increasingly frequent and prolonged episodes of palpitations over a period of 6 weeks despite a trial of bisoprolol. Echocardiography confirmed preserved systemic ventricular function, minimal atrioventricular valve regurgitation, and no evidence of outflow tract obstruction. The left atrium was not significantly dilated. Potential options for management included repeat ablation or consideration of conversion to an extracardiac Fontan.

KEYWORDS Atrial fibrillation; Atriopulmonary; Fontan; Intra-atrial reentrant tachycardia; Radiofrequency ablation
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Address reprint requests and correspondence: Dr Nicholas Jackson, Director, Electrophysiology and Pacing, Department of Cardiology, John Hunter Hospital, Lookout Rd, New Lambton Heights, Australia 2305. E-mail address: njackson193@gmail.com.

KEY TEACHING POINTS

- Patients with previous Fontan circulation surgery for treatment of complex congenital heart disease frequently present with late arrhythmias. Most typically this is intra-atrial reentrant tachycardia (IART), which may be successfully managed with catheter ablation, though consideration of Fontan revision in patients with older iterations of the procedure may be appropriate for management of refractory arrhythmia.
- Atrial fibrillation in the Fontan circulation, while less common than IART, can be effectively treated with catheter ablation using a right-sided substrate-based approach in patients with a significant scar burden, and may not require routine pulmonary vein isolation.
- This case is in keeping with other case series showing that outcomes may be favorable with ablation despite the presence of multiple IART circuits and concurrent atrial fibrillation, without the need for Fontan conversion surgery for management.

Ablation was chosen, given the satisfactory function of the Fontan circuit, systemic ventricle and atrioventricular valve, reduced periprocedural risk, and patient preference.

A 12-lead electrocardiogram of the arrhythmia is shown in Figure 1A, and is consistent with IART with variable conduction to the ventricle. Digoxin was prescribed in addition to the bisoprolol for symptom control acutely.

Prior to the procedure a cardiac computed tomography scan was performed for ablation planning (Figure 1B and C). This shows the AP Fontan circuit (in green) connecting to the right pulmonary artery, the atretic right ventricle (in red), and the pulmonary artery originating from the left ventricle and anastomosing to the aorta (Damus-Kaye-Stansel shunt).

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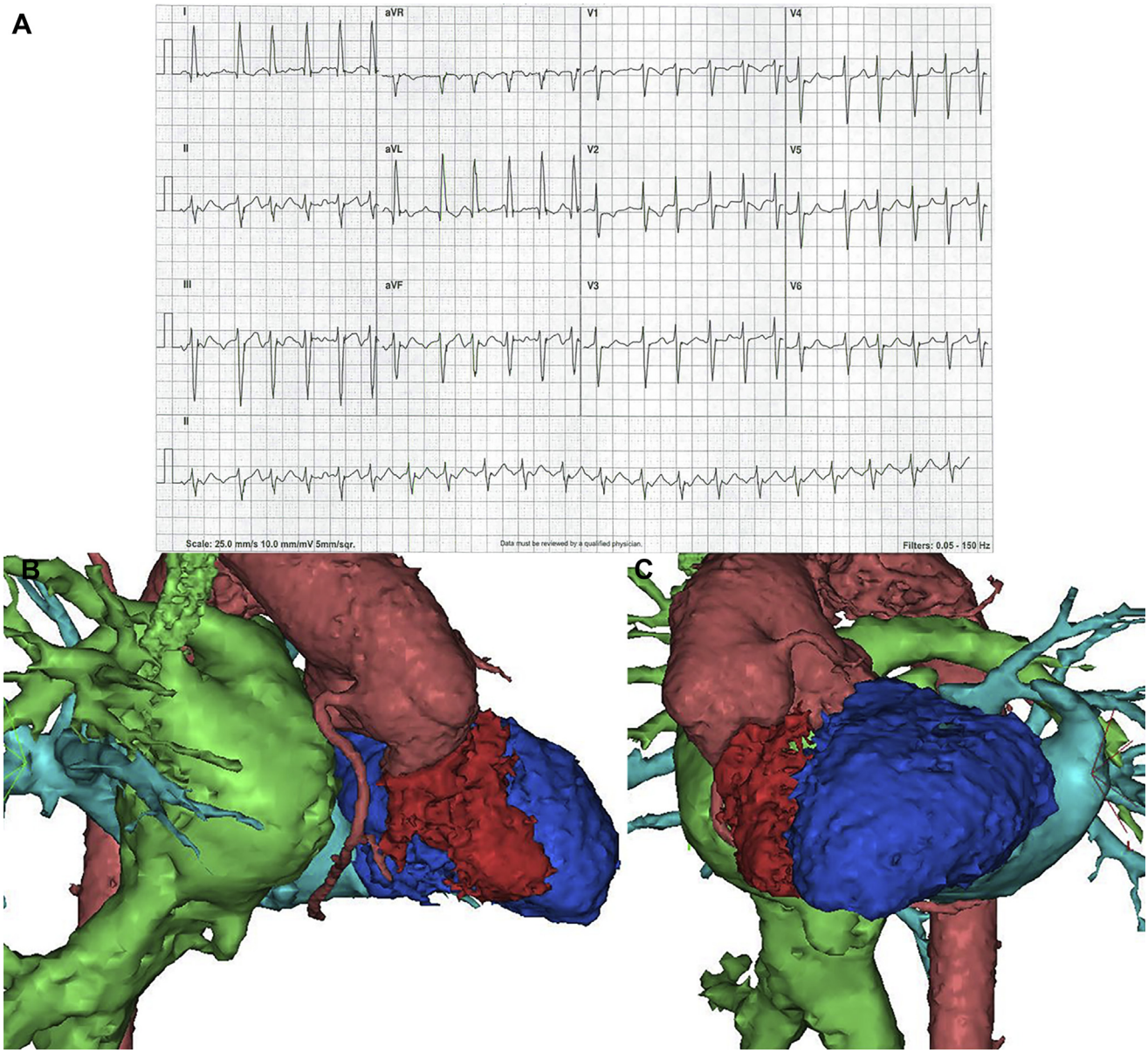

Figure 1 **A:** A 12-lead electrocardiogram of the clinical arrhythmia. **B, C:** Right anterior oblique and left anterior oblique views of the Fontan circuit (*light green*). Aorta appears in *pink*, atretic right ventricle in *red*, systemic (and morphologically left) ventricle in *dark blue*, and left atrium in *light blue*. The Damus-Kaye-Stansel connection (pulmonary artery to aorta) is seen in the left anterior oblique view.

At electrophysiology study, nonsustained atrial tachycardia was seen at baseline, and then with isoprenaline at 2 mcg/min and burst atrial pacing this tended to rapidly degenerate into AF (Figure 2A). The decision was made to perform substrate mapping of the Fontan circuit in AF, with a focus on areas with complex and fractionated signals. Mapping was performed with the CARTO 3-dimensional mapping system (Biosense Webster, Irvine, CA). The coronary sinus could not be accessed, so a duodecapolar catheter was placed around the Fontan circuit in left anterior oblique and a PentaRay catheter (Biosense Webster) was used for mapping. Fractionated electrograms predominated around the border zones of fairly extensive septal and lateral scar. Ablation was performed (35 watts) with a SmartTouch Surround Flow (Biosense Webster) irrigated ablation catheter targeting these signals, in a substrate-modification-type approach analogous to that used in persistent AF. The anterolateral and posteroseptal right atrium were the main sites targeted (as shown in Figure 2B and C), and during ablation, the AF organized into an IART (as shown in Figure 2D). Care was taken to avoid ablation at the anteroseptal region owing to the risk of heart block.

Now activation mapping of the IART could be performed (Figure 3A). This identified the entire cycle length (190 ms) of the IART on the high lateral wall of the Fontan with an area of early meeting late that demonstrated long diastolic atrial signals (Figure 3B). Ablation here then transformed activation to a second IART (Figure 3C). Activation in this

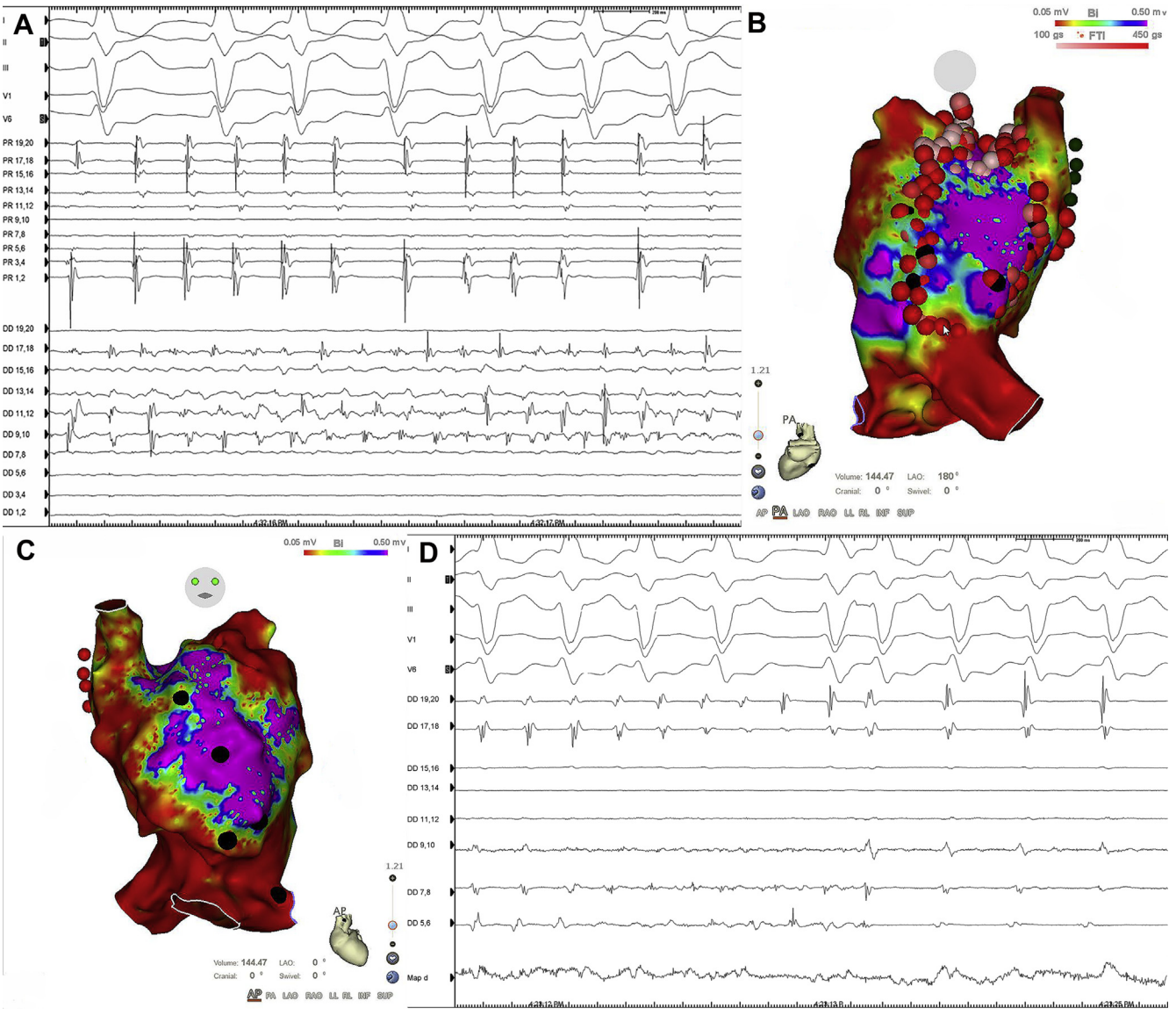

Figure 2 **A:** Atrial fibrillation. Numbered electrode pairs on a PentaRay catheter (Biosense Webster, Irvine, CA) are labeled PR, with this catheter positioned over mostly healthy tissue on the posterior wall of the Fontan circuit. Numbered electrode pairs on a duodecapolar catheter are labeled DD, with catheter orientation from proximal to distal up the anterior wall, along the roof and down the posterior wall of the Fontan circuit. **B:** Posterior view of left atrial lesion set (*red dots*). *Black dots* mark complex fractionated electrograms, with healthy voltage tissue shown in purple on this voltage map. **C:** Anterior view of the Fontan circuit. Again, healthy tissue is shown in *purple* and regions of dense scar in *red*. Ablation to organize the atrial fibrillation was not needed anteriorly. **D:** Organization of atrial fibrillation to intra-atrial reentrant tachycardia (IART) during ablation. Map d is the distal ablation catheter electrode with ablation artefact seen. Electrograms marked DD 5,6 to 9,10 show disorganized atrial activity in atrial fibrillation reverting to organized sequential activation in IART.

tachycardia was up the anterior wall and down the posterior wall through an isthmus of slow conduction at the anastomosis of the right atrium with the pulmonary artery. Ablation at the site of the isthmus led to slowing and then termination of tachycardia (Figure 3D), and nothing further could be induced with up to 3 extrastimuli down to refractoriness. The patient has not had any further symptomatic recurrence in 9 months of follow-up.

Discussion

In patients with a Fontan circulation, AF is much less common than IART[1]; however, successful ablation targeting the right atrium has been described in a limited number of cases, without requiring pulmonary vein isolation for durable success.[2–5] This patient was not known to have clinical AF prior to the ablation (although retrospective review of further electrocardiograms did reveal AF on 1 occasion). The disorganized appearance of electrograms on initial induction in this case may have represented IART with breakdown of the emanating wave fronts through the substantial scar burden in the Fontan circuit. An observational study has highlighted increasing AF with increasing age in congenital heart disease patients,[6] and the presence of previous or induced AF has recently been linked to higher recurrence rates for atrial arrhythmias in coronary

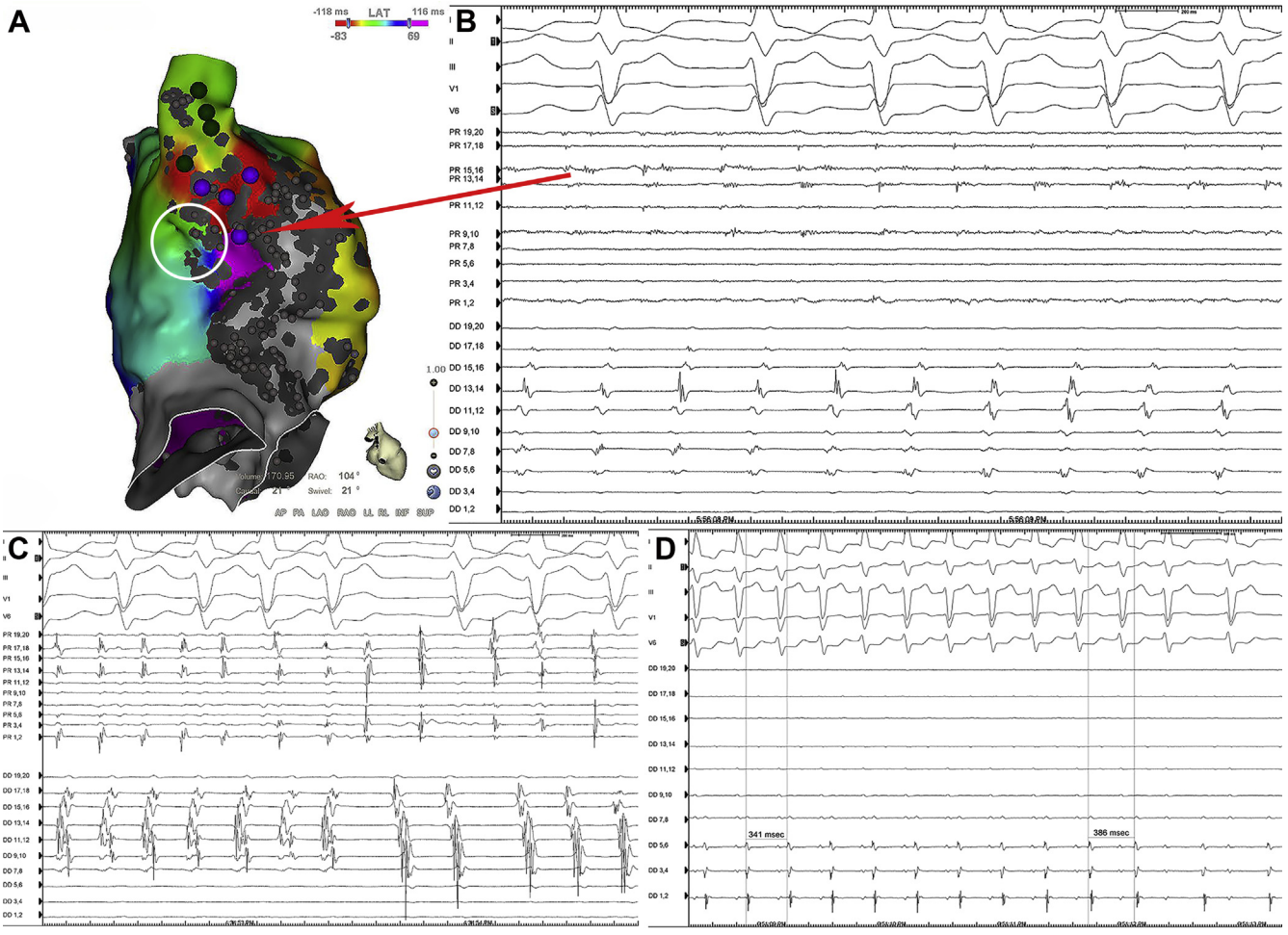

Figure 3 **A:** Right lateral view of the activation map showing the intra-atrial reentrant tachycardia (IART) circuit (*white circle with arrowheads*). Complex fractionated electrograms in IART are marked with purple dots. **B:** Complex fractionated and mid-diastolic electrograms on the PentaRay catheter capturing the entire tachycardia cycle length. The red arrow indicates the electrogram where the first IART transitioned with ablation. **C:** Transition of first IART to a second IART. Electrograms marked DD show duodecapolar catheter poles oriented with the proximal poles anteriorly, and the catheter pointing up the anterior wall, along the roof in an anteroposterior direction with distal poles against the posterior wall of the Fontan circuit. **D:** Slowing and termination of second IART to sinus rhythm.

heart disease patients undergoing catheter ablation.[7] AF may be a marker for greater scar burden in these patients and potentially indicates the need for more extensive ablation.

As is commonly seen in IART with the Fontan circulation, multiple circuits were mapped. A combined approach of substrate modification (targeting abundant complex and fractionated signals along scar borders) and activation mapping was successful in this case. In our clinical experience with IART in the Fontan circuit, entrainment attempts frequently lead to change in the tachycardia circuit or termination without additional diagnostic yield. Since the whole tachycardia cycle length was captured here and prolonged and fractionated signals were identified at the isthmus in atrial diastole, this was deemed adequate evidence of the circuit for ablation.

The choice to pursue further ablation, vs performing conversion to an extracardiac total cavopulmonary circulation, depends on multiple considerations. Although rhythm control is significantly improved with conversion (similar to the success of catheter ablation[1,8]), there is a significant periprocedural risk of 5%–10% mortality with a prolonged recovery time compared to ablation.[9–11] In observational series, Fontan conversion (when performed early) seems to be associated with better short- and long-term outcomes.[10]

This case further demonstrates that AF in patients with congenital heart disease and a significant scar burden can be successfully treated by isolating regions of scar and fractionated electrograms to organize the rhythm to IART. This patient had complex congenital heart disease with multiple congenital abnormalities, multiple prior surgeries, and 2 different IARTs as well as AF but still had an excellent clinical outcome from ablation at 9 months follow-up.

References

1. Moore BM, Anderson R, Nisbet AM, et al. Ablation of atrial arrhythmias after the atriopulmonary Fontan procedure: mechanisms of arrhythmia and outcomes. JACC Clin Electrophysiol 2018;4:1338–1346.
2. Kollengode M, Mathew J, Yeung E, Sauer WH, Nguyen DT. Successful atrial fibrillation ablation without pulmonary vein isolation utilizing focal impulse

and rotor mapping in an atriopulmonary Fontan. HeartRhythm Case Rep 2018; 4:241–246.
3. De Groot NM, Blom N, Vd Wall EE, Schalij MJ. Different mechanisms underlying consecutive, postoperative atrial tachyarrhythmias in a Fontan patient. Pacing Clin Electrophysiol 2009;32:e18–e20.
4. Ruckdeschel ES, Kay J, Sauer WH, Nguyen DT. Atrial fibrillation ablation without pulmonary vein isolation in a patient with Fontan palliation. Card Electrophysiol Clin 2016;8:161–164.
5. Takahashi K, Shoda M, Manaka T, Nakanishi T. Successful radiofrequency catheter ablation of atrial fibrillation late after modified Fontan operation. Europace 2008;10:1012–1014.
6. Labombarda F, Hamilton R, Shohoudi A, et al. Increasing prevalence of atrial fibrillation and permanent atrial arrhythmias in congenital heart disease. J Am Coll Cardiol 2017;70:857–865.
7. Roca-Luque I, Rivas-Gandara N, Dos Subira L, et al. Long-term follow-up after ablation of intra-atrial re-entrant tachycardia in patients with congenital heart disease: types and predictors of recurrence. JACC Clin Electrophysiol 2018; 4:771–780.
8. Song MK, Bae EJ, Kwon BS, et al. Intra-atrial reentrant tachycardia in adult patients after Fontan operation. Int J Cardiol 2015;187:157–163.
9. d'Udekem Y, Iyengar AJ, Galati JC, et al. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. Circulation 2014;130:S32–S38.
10. Poh CL, Cochrane A, Galati JC, et al. Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion. Eur J Cardiothorac Surg 2016;49:530–535; discussion 535.
11. Mavroudis C, Deal BJ. Fontan Conversion: Literature Review and Lessons Learned Over 20 Years. World J Pediatr Congenit Heart Surg 2016;7:192–198.