Successful Fontan conversion combined with cardiac resynchronization therapy for a case of failing Fontan circulation with ventricular dysfunction

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Abstract

Although Fontan conversion combined with cardiac resynchronization therapy appears to be an effective surgical solution for the management of failing Fontan circulation with refractory atrial arrhythmia and cardiac dysfunction due to dysynchronous ventricular wall motion, limited data are available on the mid- to long-term results of this treatment. We report our successful experience with Fontan conversion combined with cardiac resynchronization therapy in a male patient with failing Fontan circulation who showed favourable outcomes 5 years after the operation.

Keywords: Failing Fontan • Fontan conversion • Cardiac resynchronization therapy

INTRODUCTION

Failing Fontan, causing low-output syndrome, atrial arrhythmia, desaturation, systemic ventricular dysfunction, thrombus formation and protein-losing enteropathy, presents a serious problem long after Fontan surgery [1]. Although Fontan conversion combined with cardiac resynchronization therapy (CRT) appears to be an effective surgical solution for the management of failing Fontan circulation with refractory atrial arrhythmia and cardiac dysfunction, the data available on the mid- to long-term results of this treatment are limited [2, 3]. We report our successful experience with Fontan conversion combined with CRT in a patient with failing Fontan and heart failure due to ventricular dysynchrony in a functional single ventricle.

CASE

The patient was a 27-year old man with single right ventricle morphology. The details of his cardiac morphology were as follows: (A[I],X,L), double-inlet right ventricle, left-sided atroventricular valve atresia, pulmonary valve atresia, rudimentary left ventricle, left aortic arch and dextrocardia associated with right isomerism. The patient underwent a Fontan operation (atriopulmonary connection) at the age of 14 years.

After the initial Fontan operation, he showed no symptoms and was in New York Heart Association (NYHA) class I without medication until the age of 26 years. At the age of 26, the patient presented with congestive heart failure and atrial tachyarrhythmia. At that time, he was in NYHA functional class III. An X-ray showed cardiomegaly with pulmonary congestion. An electrocardiogram showed atrial tachycardia (AT) with a wide QRS wave of 162 ms and a right bundle branch block pattern. His plasma brain natriuretic peptide (BNP) level was 900pg/ml. Echocardiography showed diffuse hypokinesis of the systemic right ventricle. Cardiac catheterization showed an elevated right atrial pressure of 15 mmHg and a decrease in the oxygen saturation of the aorta to 92% due to right-to-left shunting at the atrial septum baffle leak. Right atrium (RA) angiography showed a dilated RA and right-to-left shunting between the RA and pulmonary venous chamber. Ventriculography revealed diffuse hypokinesis of the ventricle (ejection fraction of 31%) and asynchronous ventricular wall motion between the systemic right ventricle (RV) and rudimentary left ventricle (rLV) (Fig. 1A and B). After admission, cardiac support and medication were initiated using catecholamine, diuretics, beta-blockers and an angiotensin-converting enzyme inhibitor. Although catheter ablation (CA) was performed for AT, multiple ATs originating from the anatomical RA were induced, and therefore, all induced ATs could not be ablated. Low-output syndrome, a large RA with resistant AT, and right-to-left shunting through the atrial baffle leakage seemed to indicate that Fontan conversion concomitant with arrhythmia surgery was necessary, but the depressed ventricular wall motion was thought to represent a serious detrimental effect, even after Fontan conversion. A multisite pacing study performed by the transaortic insertion of a pacing catheter into the right and rudimentary left ventricle showed improvement of asynchronous ventricular wall motion to

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synchronous ventricular wall motion as detected by ventriculography (Fig. 1C and D). During early systole, the rudimentary left ventricle is in systole but the systemic right ventricle is still relaxing. After mid-systole, this association reversed. The synchrony was improved by multisite pacing (C and D). (A) Ventriculography and schema in the early-systolic phase during sinus rhythm. (B) Ventriculography and schema in the mid- to late-systolic phase during sinus rhythm. (C) Ventriculography and schema in the early-systolic phase during multisite pacing. (D) Ventriculography and schema in the mid- to late-systolic phase during multisite pacing. RV: systemic right ventricle; rLV: rudimentary left ventricle.

Figure 1: Ventriculography revealed asynchronous ventricular wall motion between the systemic right ventricle and rudimentary left ventricle during sinus rhythm (A and B). During early systole, the rudimentary left ventricle is in systole but the systemic right ventricle is still relaxing. After mid-systole, this association reversed. The synchrony was improved by multisite pacing (C and D). (A) Ventriculography and schema in the early-systolic phase during sinus rhythm. (B) Ventriculography and schema in the mid- to late-systolic phase during sinus rhythm. (C) Ventriculography and schema in the early-systolic phase during multisite pacing. (D) Ventriculography and schema in the mid- to late-systolic phase during multisite pacing. RV: systemic right ventricle; rLV: rudimentary left ventricle.

synchronous ventricular wall motion as detected by ventriculography (Fig. 1C and D). A comparison of haemodynamic data between the AAI pacing mode (pacing rate of 90 bpm) and multisite pacing (pacing rate of 90 bpm with an atrioventricular delay of 120 ms) showed that the QRS duration on the 12 lead electrocardiogram narrowed from 160 to 112 ms, pulmonary wedge pressure decreased from 10 to 6 mmHg, central venous pressure decreased from 14 to 12 mmHg and end-diastolic pressure of the systemic ventricle decreased from 6 to 4 mmHg. There was no difference between the atrial and pulmonary artery pressures. Multisite pacing raised the maximum dp/dt by 65 mmHg/s and systolic blood pressure by 5 mmHg. On the basis of these results, we scheduled a Fontan conversion combined with CRT for this 27-year old patient. Fontan conversion using a 24-mm diameter vascular graft (Gore-Tex Vascular Grafts; Gore & Associates, Phoenix, AZ, USA) with a right-sided maze procedure was performed. Since the left side of the inferior vena cava was distal to the apex on the right side, the conduit could be created without curvature. After weaning from the cardiopulmonary bypass, CRT was performed. An atrial lead (4968-35; Medtronic, Inc., Minneapolis, MN, USA) was positioned on the left side of the anatomical RA. Ventricular leads were positioned on the apex of the systemic RV (4968-35; Medtronic, Inc.) and the posteriorly located r-LV (511211 MyoPore; Greatbatch Medical, Clarence, NY, USA). Insync III (Medtronic, Inc.) was implanted and programmed in dual-chamber pacing mode (rate 80–130 bpm) with an atrioventricular delay of 120 ms and a the interventricular delay of 0 ms. An electrocardiogram showed that CRT narrowed the duration of the QRS wave from 160 to 110 ms. Because haemodynamics remained stable after the completion of the Fontan conversion with CRT, we did not create a fenestration in the conduit. During 5 years of postoperative follow-up, no AT episodes or other cardiac events occurred. The patient’s NYHA classification improved from Class III to Class I after the operation. At 5 years after the surgery, he remains in NYHA class I, his cardiothoracic rate on a chest X-ray is 45%, the plasma BNP level is 59.3 pg/ml, and the ejection fraction of the systemic ventricle on echocardiography is 48% (Fig. 2). His current medication regimen consists of diuretics, beta-blockers and an angiotensin receptor blocker.
DISCUSSION

Fontan conversion concomitant with CRT has recently become the most common surgical treatment for failing Fontan [2, 3]. Atrial arrhythmia is a major complication that reduces quality of life long after Fontan surgery. In this case, radiofrequency CA for AT was performed before the Fontan conversion. An electrophysiological study and electroanatomic mapping (CARTO; Biosense Webster) revealed the presence of a widespread electrical low-voltage area and scarring in the RA, as well as multiple inducible intra-atrial macroadreentrant ATs. We attempted to perform CA for the ATs, but complete success was not achieved. A high recurrence rate of AT after CA was expected in this case. Fontan conversion with arrhythmia surgery is reportedly efficacious for failing Fontan with refractory AT [1–3]. We performed a right-sided maze procedure at the Fontan conversion. In this case, the patient also had ventricular dysfunction in the functional single ventricle due to incoordinate ventricular wall motion between the systemic RV and rLV. Even after successful Fontan conversion, ventricular dysfunction continues to represent a serious difficulty. CRT is another choice for the management of ventricular dysfunction after a Fontan operation with single-ventricle physiology. The prolonged QRS reflected interventricular and intraventricular conduction delays, and ventriculography showed mechanical dyssynchrony of the functional single ventricle. In this case, because of the complex cardiac anatomy, a multsite pacing study was performed to determine the optimal pacing site and evaluate ventricular synchrony using ventriculography. The pacing study revealed not only a narrowed QRS duration but also an improved synchrony of ventricular contractions. On the basis of this pacing study result, we added CRT to the Fontan conversion. Although the acute effects of CRT for a single ventricle have been reported [4], the long-term effects have not been documented. In this case, the patient maintained his improved clinical status during the 5 years of follow-up. Heart transplantation is the final option reserved for a failing Fontan with advanced myocardial dysfunction, but some authors have suggested that rescue heart transplantation for a failing Fontan is unwarranted because early mortality after heart transplantation is high at ~30% [5]. Fontan conversion combined with CRT is a valid option for rescuing a failing Fontan with cardiac dysfunction associated with ventricular dyssynchrony. A further, larger study is required to establish the long-term efficacy of this treatment in this population.

Conflict of interest: none declared.

REFERENCES