Successful Fontan Completion After Cardiac Resynchronization Therapy

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This patient underwent a modified Norwood operation with a right ventricle–pulmonary artery shunt at the age of 7 days following the diagnosis of hypoplastic left heart syndrome with mitral and aortic atresia; this procedure was followed by bidirectional Glenn operation at the age of 5 months. The girl subsequently developed congestive heart failure despite the administration of angiotensin-converting enzyme inhibitors and β-blockers after the initial operation. At the age of 11 months, she required hospitalization and intravenous inotropic support. Coil embolization and surgical ligation of the aortopulmonary collaterals were performed at the age of 1 year but could not control the heart failure. The maximum plasma brain natriuretic peptide level after the second palliative operation was 3823 pg/mL. Cardiac catheterization performed at the age of 1 year and 4 months revealed ventricular dyssynchrony, ventricular ejection fraction of 36.2%, right atrial pressure of 7 mm Hg, superior vena cava pressure of 5 mm Hg, and right ventricular pressure of 35 mm Hg. The heart failure was subsequently controlled with medical treatment; at the age of 2 years and 2 months, the patient underwent successful Fontan completion with extracardiac conduit through bidirectional cavopulmonary anastomosis. The cardiac output was 4.2 L/min, systemic arterial oxygen saturation was 98%, and the maximum oxygen consumption was 250 mL/kg/min. The ECG showed complete right bundle branch block with a QRS duration of 184 ms. The left ventricular pressure was 50 mm Hg, and the mean pulmonary arterial pressure was 16 mm Hg. The color Doppler echocardiogram revealed good flow through the conduit. The right ventricular systolic pressure was 27 mm Hg, and the right atrial pressure was 5 mm Hg. The right ventricular diastolic pressure was 12 mm Hg. The patient has been doing well without heart failure since the Fontan completion.
Figure 2. Two-dimensional transthoracic echocardiographic measurements of circumferential strain before (A) and after (B) CRT. Colors of the lines correspond to those of wall segments. The bottom green line shows ECG. Ventricular dyssynchrony can be seen before CRT (early in the posterior wall and delayed in the anterior wall). After CRT, there were no regional differences in the timing of the peak strain. CRT indicates cardiac resynchronization therapy.
16 mm Hg, and grade 2 tricuspid regurgitation. QRS duration was 184 ms (Figure 1), and arterial oxygen saturation was 80%. To detect the most delayed and earliest contraction points, 2-dimensional speckle-tracking transthoracic echocardiography was performed with Philips iE33 (Philips Healthcare, Bothell, WA; Transducer: S5-1) to assess the circumferential strain by using short-axis views (Figure 2A and online-only Data Supplement Movie I), and the data were analyzed by using Philips QLAB Version X.X (Philips Healthcare, Bothell, WA). Echo evaluation identified the most delayed contraction point at the right ventricle outflow and the earliest contraction point at the posterior wall. Cardiac resynchronization therapy (CRT) with DDD pacemaker implantation and tricuspid valve plasty were performed on January 25, 2010. The precise positions of the most delayed and earliest contraction points were confirmed by epicardial echo evaluation. The most delayed contraction point was at the right ventricle outlet between the anterior delimiting artery and the previous right ventricle–pulmonary artery conduit anastomosis, and the earliest contraction site was at the hypoplastic left ventricle component. Permanent atrial pacing leads were placed on the Waterstone groove. Systemic blood pressure was assessed by atrial sensing and sequential ventricular pacing with use of temporary leads and an indwelling arterial catheter at various ventricular pacing sites. The largest increase in systemic blood pressure was achieved with simultaneous bipolar ventricular pacing set at 120 bpm and atrioventricular delay of 120 ms. Despite hemodynamic improvement, transesophageal echocardiography showed persistent mild-to-moderate tricuspid regurgitation, for which tricuspid valve plasty with interrupted suture at the medial commissure and Kay-Reed annuloplasty at the anteroposterior commissure were performed. Following the establishment of biventricular pacing, the patient was successfully weaned from cardiopulmonary bypass. Cardiac catheterization performed 3 months later revealed decreased superior vena cava pressure (14 mm Hg), increased ejection fraction (43.9%), and reduced dyssynchrony (Figure 2B and online-only Data Supplement Movie II) and tricuspid regurgitation, in comparison with the preoperative findings. QRS duration was 112 ms (Figure 3), and brain natriuretic peptide level was decreased to 443 pg/mL. One month after, the patient underwent Fontan operation. Her postoperative course was uneventful 1 year later, and cardiac function had further improved with an ejection fraction of 57.6%, QRS duration of 106 ms, and brain natriuretic peptide level of 42.4 pg/mL.

CRT is a good option for patients with heart failure caused by cardiac dyssynchrony. Determination of the pacing site is most important for the efficacy of CRT. In the present case, intraoperative 2-dimensional epicardial echocardiographic measurement of circumferential strain enabled us to precisely identify the most delayed and earliest contraction points. We could also evaluate the increase in systemic blood pressure and the improvement in synchrony on a real-time basis. Almost all previous studies reporting successful CRT used multisite ventricular pacing with a CRT pacemaker. Considering the body size of our patient, we chose a DDD pacemaker as the CRT device. The DDD pacemaker could not control ventriculoventricular delay but could ameliorate severe heart

Figure 3. Postoperative ECG showing a decreased QRS duration of 112 ms following ventricular pacing (DDD mode).
failure when the pacing leads were placed at optimal sites. Our case showed not only successful completion of Fontan operation, but also further improvement of cardiac function 1 year after the Fontan operation. CRT may be an effective treatment option for patients with a functional single ventricle who develop heart failure, and it may result in long-term benefits in these patients.

Disclosures

None.

References

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