Feasibility of Cardiac Resynchronization Therapy in a Patient with Complex Congenital Heart Disease and Dextrocardia, Facilitated by Cardiac Computed Tomography and Coronary Sinus Venography

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We describe a case with pacemaker implantation for cardiac resynchronization therapy (CRT) in a patient with complex congenital heart disease, facilitated by cardiac computed tomography (CT) and coronary sinus (CS) venography. A 37-year-old male presented with congenitally corrected transposition of the great arteries and mesocardia, along with a history of two open heart surgeries (closure of atrial septal defects and a ventricular septal defect, and pulmonary valvectomy at age 7; mechanical tricuspid valve replacement at age 13). He showed symptoms of progressive heart failure (NYHA class III) with significant impairment of the systemic right ventricular function. He also developed permanent atrial fibrillation with a junctional rhythm at a rate of 45 beats per minute. Biventricular pacing without an atrial lead was considered to be the best option available. CRT implantation was facilitated by proper identification of CS anatomy utilizing cardiac CT and CS venography and was performed without any complications. At follow up, a postero-anterior chest X-ray showed the final position of the right-sided ventricular (left ventricular morphology) lead pointing to the apex and the left ventricular lead at the posterolateral aspect of the systemic ventricle (right ventricular morphology).

Adult patients with congenital heart disease represent an expanding population that is growing at a rate of 5% per annum.1 Ventricular dysfunction is common in adult congenital heart disease patients who have a systemic right ventricle (RV) and is related to electromechanical dyssynchrony.2-4 Over 25% of such individuals ultimately progress to symptomatic heart failure, which is occasionally refractory to drug therapy and is associated with substantial morbidity and mortality.4 Therefore, identification of novel therapeutic strategies in these patients is of critical importance.

Permanent pacemaker leads may be implanted either epicardially or transvenously in infants as well as older children.5 Patients with a systemic RV may require conventional pacemaker therapy,6 which in the presence of ventricular dysfunction and conduction disease may further compromise cardiac performance.7,8 Hence, devices capable of combining conventional pacing with modern functions geared towards ameliorating ventricular function seem to offer an obvious advantage.

Although cardiac resynchronization therapy (CRT) is rapidly emerging as an effective strategy for managing ventricular...
dysfunction and heart failure associated with congenital heart disease, its usage in these patients is limited in terms of patient numbers and follow up. Experience in transvenous CRT implantation is limited further by distorted coronary sinus (CS) anatomy and its tributaries due to previous surgeries and intractable heart failure. However, this route may be feasible for future lead implantation at a greater age.

In this report, we described the feasibility of transvenous CRT implantation in a patient with congenitally corrected transposition of the great arteries (ccTGA) and mesocardia, along with a history of two open heart surgeries.

Case presentation

A 37-year-old male presented with ccTGA and mesocardia, along with a history of two open heart surgeries. At the age of seven years, he underwent surgical closure of atrial septal defects and a ventricular septal defect, and pulmonary valvectomy. He also underwent mechanical tricuspid valve/systemic valve (TV) replacement at the age of 13 years. On follow up he showed symptoms of progressive heart failure with significant impairment of systemic RV (RV morphology) function. He also developed permanent atrial fibrillation with junctional rhythm at a rate of 45 beats per minute. In view of the slow ventricular response and concomitant severe heart failure (NYHA class III) symptoms, biventricular pacing without an atrial lead was considered to be the best option available. It was anticipated that the abnormal CS anatomy would be more difficult in this particular patient, hence cardiac computed tomographic (CT) angiography was performed as a road to implantation procedure (Figure 1).

CRT was performed as a two-step procedure in a single session. Firstly, CS angiography was performed through the right femoral vein approach using a pre-shaped long sheath (St. Jude Medical, SL3) and occlusive balloon (Medtronic, Attain) to plan for proper systemic ventricular lead positioning (Figure 2). Occlusive venography showed disturbed CS anatomy with only one suitable posterolateral branch, as well as the incidental finding of a fistula connecting the CS to the left atrium (Figure 2).

Subsequently, two axillary vein accesses were obtained, and then the RV lead (Medtronic, Sprint Quatro, 6947) was implanted in the right-sided ventricular apex (LV morphology). The LV delivery system (9F safe sheath CS Guide) was introduced into right

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**Figure 1.** An axial computed tomography angiographic view depicting (A) artificial tricuspid valve (TV), right atrium (RA), inferior vena cava (IVC), small fistula into the left atrium (LA). In the three dimensional reconstruction (B) the coronary sinus is color coded in blue. DAO – descending aorta; LV – left ventricle; RV – right ventricle.
atrium, then into the CS with the help of a 6F Judkins right catheter and 0.035" terumo guide wire (Figure 3). Another CS venography was obtained through the safe sheath to rule out any dissection and to ensure that the sheath was in the true lumen (Figure 4). A bipolar 4194 LV pacing lead (Medtronic) was then introduced into the target vein without much difficulty. The SL3 with the Attain balloon were pulled back through the femoral vein before the safe sheath was withdrawn under fluoroscopy guidance (Figure 5).

The procedure was then completed without any complications. At follow up, the postero-anterior chest X-ray showed the final position of the right-sided ventricular (LV morphology) lead pointing to the apex and the LV lead at the postero-lateral aspect of the systemic ventricle (RV morphology) (Figure 6).

Discussion

Congenitally corrected transposition of the great arteries accounts for 0.5% of all cases of congenital heart disease, of which 30% might be associated with mesocardia. Additional anomalies may be present, such as ventricular septal defect in 60-80%, sub-pulmonary ventricle outflow obstruction in 30-50% and TV abnormalities in 14-56% of patients. Rhythm disturbances can develop in up to 2% of this population per year, with or without surgical intervention. In most of these cases, failure of the sub-aortic ventricle usually appears in the third decade of life, with or without corrective or palliative surgeries.

Although pacemaker implantation is considered...
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the best option for dyssynchrony in ccTGA patients, the optimal pacing mode and pacing site in this group of patients are not well established. There is also evidence to show worsening of heart failure after conventional dual-chamber pacing with the ventricular lead placed in the sub-pulmonary ventricle in ccTGA patients.

CRT appears to be an attractive option for ccTGA patients. In unselected populations with a systemic RV, 4-9% of patients are potentially eligible for CRT. However, deleterious effects of unintentional pacing from the middle cardiac vein have been reported in patients with ccTGA. The CS is situated adjacent to the systemic RV and drains into the systemic right atrium in ccTGA patients, which makes transvenous CRT feasible in this group of patients, where it is known to improve acute hemodynamics. Because of the heterogeneous venous anatomy, visualization of the CS anatomy before attempting CRT implantation is strongly recommended, using different modalities such as CS venography, cardiac CT and magnetic resonance imaging. In addition, two-dimensional strain echocardiography is a reliable tool for detecting RV dysfunction.

Our case highlights the feasibility of transvenous CRT implantation in a patient with a complex congenital heart disease, involving a systemic sub-aortic right ventricle, provided the procedure is facilitated by proper identification of coronary sinus anatomy utilizing cardiac CT and CS venography. The presence of mesocardia in some of these cases should not exclude them from such beneficial therapies.

Figure 4. Left anterior-oblique view at 20°, coronary sinus venography from above using a 9F safesheath coronary sinus guide (CSG) with one suitable posterolateral branch (black arrow).

Figure 5. Left anterio-oblique view at 20° showing lead (Medtronic 4194) in the posterolateral position (black arrow) emerging from the safe sheath, which was still in the coronary sinus, and the Attain balloon pulled back from the femoral vein (white arrow).

Figure 6. Postero-anterior chest X-ray with the final position of the right-sided ventricular (left ventricular morphology) lead pointing to the apex (white arrow), and the left ventricular lead at the posterolateral aspect of the systemic ventricle (right ventricular morphology) (black arrow).
References