VIA MEDICA

CASE REPORT

Cardiology Journal
2010, Vol. 17, No. 5, pp. 503–508

ISSN 1897–5593

Resynchronization therapy transvenous approach in dextrocardia and congenitally corrected transposition of great arteries

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Abstract

Cardiac resynchronization therapy (CRT) is an acknowledged treatment for advanced heart failure in acquired dilated cardiomyopathy, resistant to pharmacotherapy. Although there are no therapeutic standards regarding heart failure originating from congenital heart defects with systemic right ventricle, a number of CRT implantations by transvenous approach in congenitally corrected transposition of the great arteries (CCTGA) have been reported since 2001, even though none of them expressly referred to a case concomitant with dextrocardia and situs inversus anomaly.

We present a 57 year-old patient with dextrocardia and CCTGA, who underwent surgical closure of interatrial and interventricular septal defects at the age of 19, and in whom a VVI pacemaker was subsequently implanted at age 36. A three-lead CRT system was implanted by transvenous approach. Imaging techniques, including multi-slice computed tomography, targeted to pacing system and unusual anatomical relationships were applied. Within a 20-month follow-up, a significant improvement of functional NYHA class, systemic right ventricle ejection fraction and exercise capability were observed.

Entirely transvenous CRT system implantation is feasible in patients with dextrocardia and CCTGA, and has substantial potential for long-term benefits. (Cardiol J 2010; 17, 5: 503–508)

Key words: cardiac resynchronization therapy, CCTGA, transvenous approach, dextrocardia in situs viscerum inversus, multi-slice computed tomography

Introduction

Biventricular pacing in dilated cardiomyopathy with severe left ventricular (LV) systolic dysfunction, electromechanical delay and chronic heart failure (HF), despite optimal pharmacotherapy, has been demonstrated in randomized clinical studies to improve overall quality of life, functional class, exercise capability and echocardiographic parameters [1, 2]. Cardiac resynchronization therapy

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Received: 9.07.2009 Accepted: 18.10.2009

(CRT) has recently also been proved helpful in reducing all-cause mortality or unplanned hospitalization for major cardiovascular events [3, 4].

Despite no established clinical standards for CRT in congenital heart disorders, such interventions are still being embraced in advanced HF cases in adults and children alike [5–8]. It must be emphasized that CRT is widely acknowledged to be the most challenging among heart pacing procedures, especially in terms of the placement of the lead for LV stimulation, using the transvenous approach through the coronary sinus.

An additional difficulty lies in the variable anatomy of the coronary sinus itself, this being especially true for any congenital disorders. Dextrocardia complicates the procedure even more, because the shapes of commercially available LV pacing devices are suitable for left-hand sided coronary sinus in the left-subclavian vein approach [9].

In congenital corrected transposition of great arteries (CCTGA), the anatomical LV connects the right atrium to a posteriorly placed pulmonary artery. The anatomical right ventricle (RV) connects the left atrium to the anteriorly placed aorta. Owing to the abnormal relationship between the atria and the ventricles, and the ventricles with the aorta and the pulmonary artery respectively, we are actually looking at a functional correction of the circulation. Consequently, blood flows in the normal direction, although passing through the "wrong" ventricular chamber [10].

The atrio-ventricular (AV) node and His bundle have an unusual position and course, thus making the conduction system inverted, whereas His bundle is elongated. Many patients also reveal dual AV nodes. The second anomalous AV node and bundle are usually anterior, whereas the penetrating bundle is vulnerable to fibrosis with advancing age. This makes the conduction system tenuous, with a progressive incidence of complete AV block occurring at ≈2% annually [10, 11]. It should be noted at this point that ventricular septal defect (VSD) surgery, as in this particular case, may also precipitate heart block. It makes it necessary to perform long-term pacing with negative hemodynamic effects on the systemic RV which constitutes first risk factor for gradual development of HF.

The anatomy of coronary arteries is concordant with the morphology of ventricles. Systemic RV is perfused only by a single right coronary artery, which, through exposure to systemic pressure, may cause substantial impairment to myocardial perfusion [10, 12, 13]. This constitutes a second risk factor for HF in those patients. Most of them survive until advanced age and become good candidates for CRT therapy [5, 6].

The coronary sinus is ontogenically a part of the left atrium, being in fact located at the side of the left atrium, both in terms of morphological and circulatory constraints. Hence, the coronary sinus is adjacent to the systemic RV in CCTGA patients. Therefore, in CCTGA, the coronary sinus and its tributaries drain blood predominantly from the systemic RV, which makes them perfectly suitable for the placement of a lead for systemic RV stimulation [5–8].

To the best of our knowledge, no cases of fully transvenous CRT system introduction via the left subclavian vein approach for the treatment of HF in patients with CCTGA associated with dextrocardia have been reported to date.

Case report

A 57 year-old man with CCTGA and situs viscerum inversus with dextrocardia had experienced several hospital admissions due to NYHA class III HF symptoms, despite optimal pharmacotherapy (carvedilol, spironolacton and cilazapril).

At the age of 19, he underwent surgery of the interatrial and interventricular septal defect (ASD and VSD) closure, complicated by the right bundle branch block.

Seventeen years later, a single ventricular chamber pacing device (VVI) with epicardial ventricular lead, introduced by surgical thoracotomy method, was implanted, due to the development of complete AV block. The lead was located in the pulmonic LV apex, whereas the pacemaker was in the epigastric region.

The patient remained asymptomatic until the age of 47, when he started experiencing dyspnoea, fatigue and paroxysmal atrial tachycardia, resulting in hospitalization twice a year on average.

At the last admission, his blood pressure was 90/60–100/70 mm Hg, whereas the pacemaker function remained normal. Ventricular pacing 70/min with the rate response rise up to 110/min, and QRS of 170 ms width were registered (Fig. 1B).

Echocardiography revealed severe systolic ventricular dysfunction, bi-atrial and bi-ventricular dilatation, with a systemic RV ejection fraction of only 24%, as assessed by single-plane Simpson rule (Table 1). Moderate anatomical mitral and tricuspid valve regurgitation, pulmonary valvular stenosis with a gradient of 40–44 mm Hg on Doppler echocardiography, and the pulmonary trunk aneurysm were also found. An interventricular mecha-

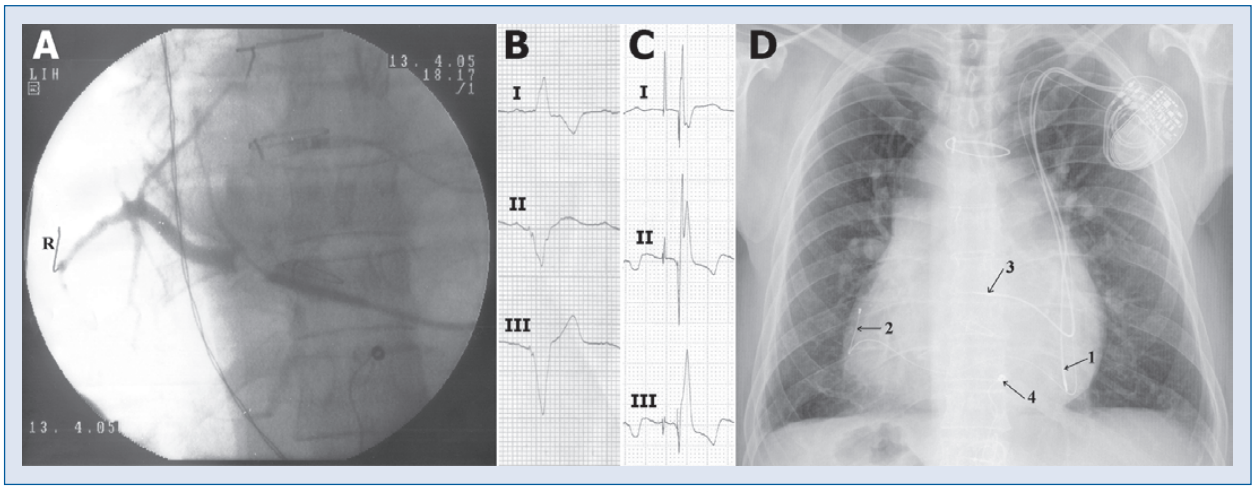

Figure 1. A. Angiography demonstrating coronary sinus with cardiac veins. The retouch of coronary sinus distal part — anterior cardiac vein (R); **B, C.** Electrocardiography. The leads I, II, III. The lead configuration adopted for dextrocardia. Paper speed 25 mm/s; **B.** Epicardial VVI pacing from the pulmonic left ventricular apex; **C.** DDD BiV (CRT), sequential atrio-biventricular pacing. Details regarding A-V and V-V delays in the text; **D.** Chest X-ray — postero-anterior projection. Three transvenous pacemaker leads are visible in the right atrium (1), the cardiac vein (2) and in the pulmonic left ventricle (3). The old, detached epicardial lead is also visible (4).

Table 1. 20-month follow-up.

	Before implantation	After implantation			
		1 month	6 months	12 months	20 months
NYHA	III	III	III	II	II
Systemic RVEF (%)	24	30	30	30	40
6-min walking test [m]	392	443	484	498	438*
Spiroergometric test (VO_2max) [mL/kg/min]	10.9	–	–	17.1	–

*Test carried out during the discopathy-originated pain symptoms; RVEF — right ventricular ejection fraction

nical dyssynchrony of 84 ms was calculated and the post-systolic contraction of the systemic RV free wall was also observed (Fig. 2A, B). The 6-min walking-test (6MWT) and spirometry revealed significant impairment of physical effort capacity (Table 1).

Coronary angiography revealed atypical coronary aortic inlets without coronary artery stenosis, i.e. the right artery and left anterior descendant artery from anterior Valsalva sinus, circumflex artery from posterior Valsalva sinus.

CRT system implantation — procedure description

Coronary sinus lumen visualization was obtained using the Medtronic Attain set (Fig. 1A).

A Medtronic 4193 stylet-driven lead was advanced into the anterior branch of the coronary sinus in order to pace the systemic RV. All previous attempts aimed at placing the lead in the posterior and postero-lateral branches of the coronary sinus had failed due to mechanical instability and high pacing thresholds. The Medtronic 5076 screw-in lead was placed in pulmonic LV outflow track underneath the pulmonary valve. Subsequently, the Medtronic 5076 screw-in lead was located in the right atrium, close to the coronary sinus outflow. These three leads were attached to a Medtronic pacemaker Insync III 8042 (Fig. 1D). The pacing and measurement of bioelectrical parameters at all the leads appeared satisfactory. Immediately after the introduction of CRT pacing, the duration of QRS has decreased from the initial 170 ms down to 140 ms (Fig. 1C).

Aiming to obtain the shortest possible interventricular dyssynchrony and optimal mitral flow time (as measured by ECHO), the atrio-ventricular and inter-ventricular delays were programmed as follows: AV delay sensed at 110 ms, AV delay

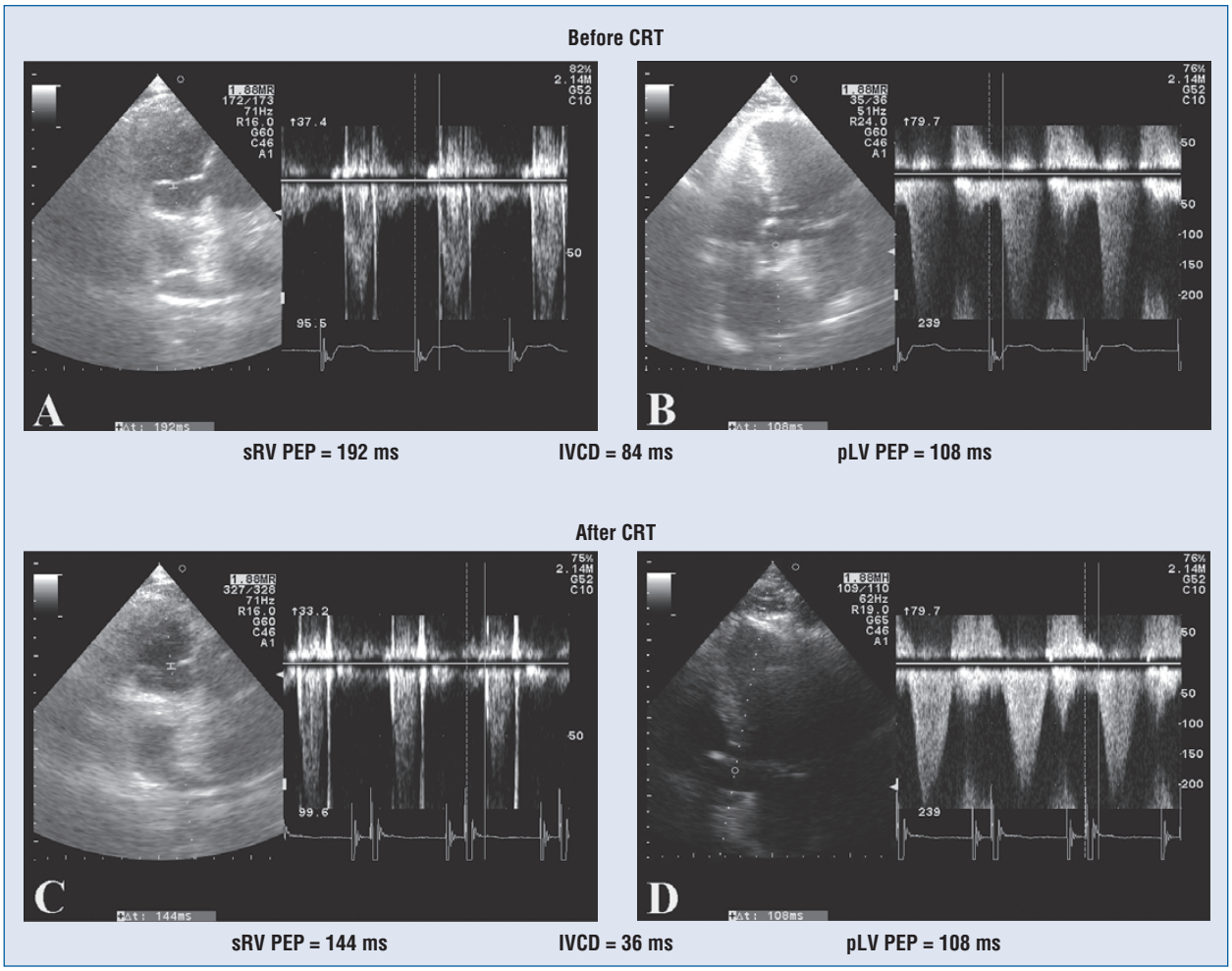

Figure 2. Echocardiography; **A, C.** Right-sided parasternal modified projection; **B, D.** Five chamber apical projection. Inter-ventricular dyssynchrony features are shown. The interventricular conduction delay (IVCD) measurement in the text; sRV — systemic right ventricle; pLV — pulmonic left ventricle; CRT — cardiac resynchronization therapy.

paced at 130 ms, and VV delay at 16 ms, with prior systemic RV stimulation. The interventricular conduction delay of 36 ms was obtained, indicating the interventricular dyssynchrony to have decreased by more than 50% in comparison to the initial value (Fig. 2C, D). The previously installed pacemaker was removed, although its 19 year-old epicardial lead was left in place.

Subsequently, the patient was discharged from hospital (presenting symptoms of NYHA class III HF) and put on cilazapril, carvedilol, spironolacton and aspirin.

20-month follow up

The follow-up was carried out one, six, 12 and 20 months following the procedure. The results of clinical observation, as assessed by NYHA class, 6MWT, spiroergometric test and systemic RV ejection fraction ECHO measurement, can be seen in Table 1.

Within one month of the procedure, atrial flutter was observed. Following initial pharmacotherapy with digoxin and an anticoagulation agent, it was converted (using rapid atrial over-drive pacing) into atrial tachycardia 170/min with the 2:1 ventricular pacing. This arrhythmia, still observed during the follow-up visit after six months, spontaneously disappeared after another six months and the sinus rhythm was re-established. Holter ambulatory recording obtained 20 months after the implantation revealed paroxysmal atrial flutter, despite the continued antiarrhythmic treatment.

MSCT visualisation

Prior to the CRT procedure, coronary angiography was performed. Multi-slice computed tomography (MSCT) was performed during the follow-up period in order to visualize the leads, arteries and venous inflow system (Fig. 3). Aorta arose from the

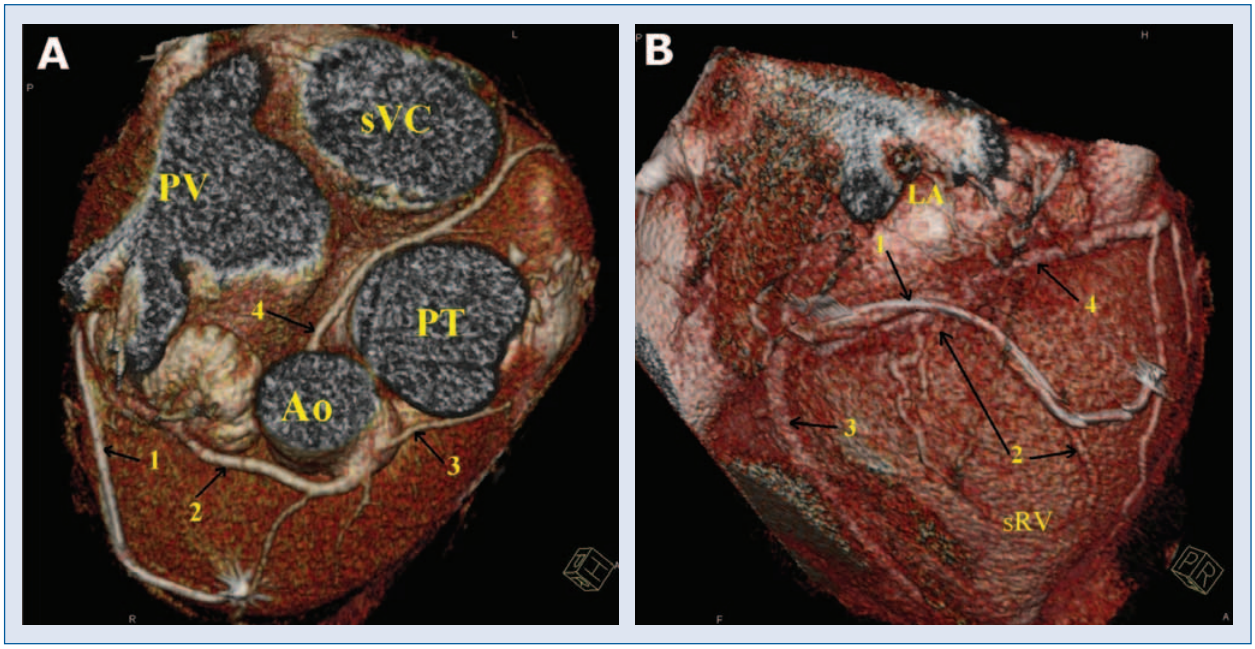

Figure 3. A, B. Multi-slice computed tomography — 3D projection. Coronary arteries and cardiac veins are shown; **A.** View of the upper part of the heart. Cut-off at the level of great vessels; 1 — coronary sinus with the lead; 2 — right coronary artery; 3 — left anterior descendent coronary artery; 4 — left circumflex coronary artery; Ao — aorta; PT — pulmonary trunk; PV — pulmonary veins; sVC — superior vena cava; **B.** View of the right lateral wall of the heart; 1 — coronary sinus with the lead; 2 — posterior and lateral branches of coronary sinus; 3 — posterior cardiac vein; 4 — right coronary artery; sRV — systemic right ventricle; LA — left atrium.

morphological RV and pulmonary trunk from the morphological LV.

Abnormal coronary artery outflows were recognized, i.e. the right artery and anterior descendent coronary artery originating from the anterior Valsava sinus, and circumflex artery originating from the posterior sinus. The right coronary artery supplied blood to the myocardium of systemic RV through a number of branches. The left anterior descendent artery (originating from the anterior Valsalva sinus in close proximity to the right coronary artery, running its course through the interventricular groove) supplies blood into the interventricular septum. The circumflex artery, turned behind the pulmonary trunk and running along the groove between the right atrium and pulmonic LV, supplied blood to the myocardium of this ventricle (Fig. 3).

The cardiac veins are also shown (Fig. 3). Coronary sinus and posterior cardiac vein circulated the blood back to the right atrium. Two veins (i.e. posterior and postero-lateral) had inlets to the coronary sinus, whereas the remnant structure resembled the anterior vein. In this vessel, the tip of systemic RV lead was found to approach the lateral systemic RV wall and then turn upward to the RV outflow track.

Discussion

Patients with CCTGA are significantly susceptible to systemic ventricle insufficiency. Key predisposing factors are the chamber of RV-like structure and blood perfusion being exposed to high blood pressure and concomitant AV conduction disturbances in the form of bundle branch blocks, up to the second and third degree AV block resulting in ventricular bradycardia. The pattern of bundle branch blocks originating from pulmonic LV pacing may generate typical pacing-induced interventricular dyssynchrony, with subsequent intensive dyssynchrony of mechanical contraction [5, 10–13].

In the study by Diller at al. [5], the demand for CRT in patients with systemic ventricle was estimated as around 4%, in due consideration of standard indications for CRT implantation.

Nowadays, the vast majority of CRT devices are implanted through the transvenous approach, widely acknowledged to be a much safer method than thoracotomy, although only a few reports deal with the actual implantation of transvenous pacing leads in CCTGA patients [6–8].

In the presently discussed CCTGA case, the additional impediment in advancing transvenous

leads consisted in situs inversus with dextrocardia in the right-handed patient, in whom the left-sided approach was preferable due to his high physical activity and possible damage to the leads in the 'crush syndrome' during active use of his right hand.

Only a single case of CRT implantation in a patient with dextrocardia was actually found in the literature, although this dealt with the right-sided transvenous approach only [9].

We effectively managed to create a three-lead stimulation system through the successful implantation of the first lead onto the antero-lateral wall underneath the aortic valve ring in conjunction with the placement of another one in the pulmonic ventricle outflow track on the anterior wall. It was not optimal in terms of theoretic rules of CRT implantation. Optimal implantation site in coronary sinus branch was attempted, but finally abandoned due to mechanical instability and high pacing thresholds. The third lead in the right atrium, in the proximity of the inflow to the coronary sinus, was also implanted. The AV and VV delays were programmed in compliance with the actual echocardiographic findings. Mechanical contraction dyssynchrony diminished by over 50%.

In the long-term, 20-month follow-up, improvement of functional NYHA class, systemic RV ejection fraction, and capacity of physical effort were noted, with no significant decrease in either of the systemic ventricle diameters.

Application of MSCT after the surgical intervention facilitated the imaging of the venous vascular bed, therefore affording a much deeper insight into the anatomical complexities of this rather rare congenital heart disorder. Coronary angiography was performed prior to the CRT procedure to ease visualisation of potential significant stenosis of coronary arteries caused by atherosclerosis in a 57 year-old patient.

Conclusions

Resynchronization therapy by a transvenous approach in patients with complex heart anatomy in CCTGA and situs inversus with dextrocardia is a feasible, low-risk surgical procedure, despite there being no established clinical standards and scarcity of pertinent experience.

Acknowledgements

The authors do not report any conflict of interest regarding this work.

No support in the form of grants, equipment or drugs has been received during the medical management of this patient and preparation of this paper.

References

1. Linde C, Leclerq C, Rex S et al. On behalf of the Multisite Stimulation In Cardiomyopathies (MUSTIC) Study Group: Long Term Benefits of Biventricular Pacing in Congestive Heart Failure: Results From The Multisite Stimulation In Cardiomyopathy (MUSTIC) Study. J Am Coll Cardiol, 2002; 40: 111–118.
2. Abraham WT, Fisher WG, Smith AL et al. For the MIRACLE Study Group: Cardiac Resynchronisation in Chronic Heart Failure. N Engl J Med, 2002; 346: 1845–1853.
3. Bristow MR, Saxon LA, Boehmer J et al. For the Comparison of Medical Therapy, Pacing, and Defibrillation in Heart Failure (COMPANION) investigators: Cardiac Resynchronisation Therapy with or without an Implantable Defibrillator in Advanced Chronic Heart Failure. N Engl J Med, 2004; 350: 2140–2150.
4. Cleland JG, Daubert JC, Erdmann E et al. Longer-term effects of cardiac resynchronization therapy on mortality in heart failure [the Cardiac Resynchronization-Heart Failure (CARE-HF) trial extension phase]. Eur Heart J, 2006; 27: 1928–1932.
5. Diller G-P, Okonko D, Uebing A, Yen Ho S, Gatzoulis MA. Cardiac resynchronisation therapy for adult congenital heart disease patients with a systemic right ventricle: Analysis of feasibility and review of early experience. Europace, 2006; 8: 267–272.
6. Janousek J, Tomek V, Chaloupecky VA et al. Cardiac resynchronisation therapy: A novel adjunct to the treatment and prevention of systemic right ventricular failure. J Am Coll Cardiol, 2004; 44: 1927–1231.
7. Krishnan K, Avramovitch NA, Kim MH, Trohman RG. Cardiac resynchronisation therapy: A potential option for congenitally corrected transposition of the great vessels. J Heart Lung Transplant, 2005; 24: 2293–2296.
8. Rodriguez-Cruz E, Karpawich PP, Lieberman RA, Tantengco MV. Biventricular pacing as alternative therapy for dilated cardiomyopathy associated with congenital heart disease. PACE, 2001; 24: 235–237.
9. Belotti G, Piti A, Curnis A. Biventricular implantable cardiac defibrillator in dextrocardia with situs viscerum inversus. Ital Heart J, 2004; 5: 559–562.
10. Warnes CA. Transposition of the great arteries. Circulation, 2006; 114: 2699–2709.
11. Fischbach PS, Law IH, Serwer GS. Congenitally corrected L-transposition of the great arteries: abnormalities of atrioventricular conduction. Prog Pediatr Cardiol, 1999; 10: 37–43.
12. Graham TPJr, Bernard YD, Mellen BG et al. Long-term outcome in congenitally corrected transposition of the great arteries: A multi-institutional study. J Am Coll Cardiol 2000; 36: 255–261.
13. Hornung TS, Bernard EJ, Celermajer DS et al. Right ventricular dysfunction in congenitally corrected transposition of the great arteries. Am J Cardiol, 1999; 84: 1116–1119.