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CASE REPORT

Successful pregnancies after transvenous cardiac resynchronization therapy in a woman with congenitally corrected transposition of the great arteries

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ABSTRACT

Congenitally corrected transposition of the great arteries is a rare heart defect that can be associated with systemic ventricular dysfunction and conduction disturbances. The use of cardiac resynchronization therapy in patients with congenital heart disease is not fully established, and achievement of successful pregnancies after implantation of transvenous, biventricular system has never been described, and which resulted in a significant clinical improvement.

We describe a 33-year-old female with congenitally corrected transposition of the great arteries, who achieved six pregnancies and successful vaginal deliveries. The two last pregnancies were achieved after cardiac resynchronization therapy for systemic ventricular dysfunction and complete heart block. A congenital cardiac disease has been identified in only one offspring.

1. Introduction

Congenitally corrected transposition of the great arteries (ccTGA) accounting for less than 1% of congenital heart disease.[1] Most women with ccTGA reach childbearing age. The morphologic right ventricle (RV) is not designed to support systemic circulation and particularly increased cardiac output during pregnancy and is predisposed to dysfunction.[2–4]

Cardiac resynchronization therapy (CRT) is supposed to stabilize if not to improve cardiac function in these patients.[5–7]

To our knowledge this is the first case reported of cardiac resynchronization therapy by transvenous access in a patient with congenitally corrected transposition of the great arteries with complete atrioventricular block (AVB) and systemic ventricular dysfunction who achieved six successful pregnancies and vaginal deliveries, four before and two after cardiac resynchronization therapy.

2. Case report

We report a case of a 33-year-old female with congenitally corrected transposition of the great arteries, who was admitted to our institution because of an exacerbation of heart failure (NYHA class III heart failure symptoms), recurrent dizzy spells and general physical weakness.

At the age of 22, she underwent surgery of an ostium primum atrial septal defect (ASD) closure complicated by a right bundle branch block. Initial postoperative atrioventricular conduction was normal. No anatomical repair with double switch operation was performed. She has also the history of 4 pregnancies and successful vaginal deliveries. The patient remained asymptomatic until the age of 33, when she started experiencing dyspnea, fatigue and dizzy spells.

At the admission, her blood pressure was 90/60 mmHg, and the heart rate was 42 bpm. Electrocardiogram (ECG) upon admission revealed complete atrioventricular block (AVB) and a relatively wide QRS escape rhythm with QRS duration of 115 ms (Fig. 1). Echocardiography revealed severe systolic ventricular dysfunction, right ventricular (RV) dilation with systematic RV ejection fraction

Abbreviations: AV, atrio-ventricular; AVB, atrioventricular block; ASD, atrial septal defect; ccTGA, congenitally corrected transposition of the great arteries; CRT, cardiac resynchronization therapy; CS, coronary sinus; LV, left ventricle; PLCV, posterolateral cardiac vein; RV, right ventricle; RVEF, right ventricle ejection fraction.

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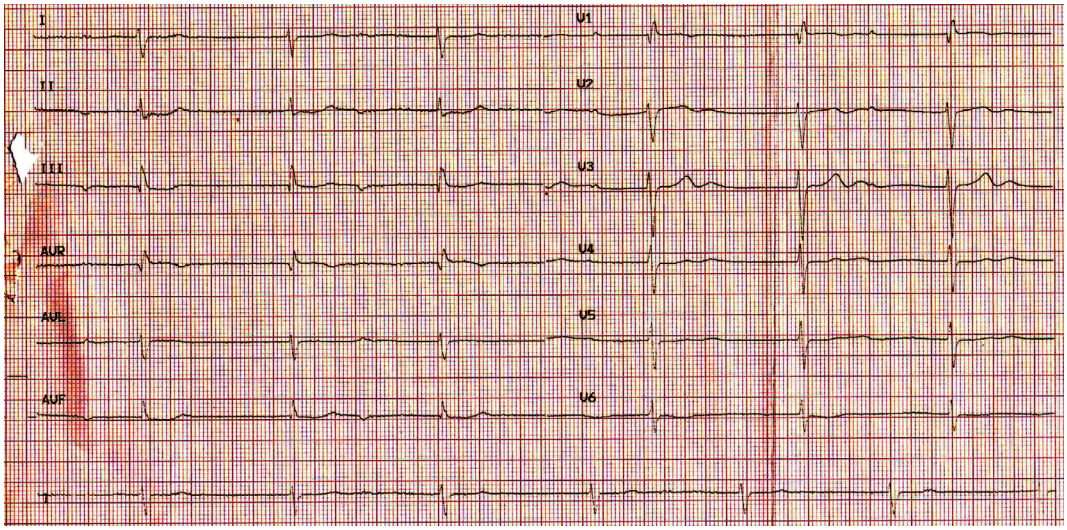

Fig. 1. Electrocardiogram (ECG) on admission showing complete heart block with a relatively wide QRS complex escape rhythm.

of only 33% assessed by single plane Simpson rule. Significant tricuspid valve regurgitation was also observed. Indexes of interventricular, and intraventricular dyssynchrony were not studied.

Given the underlying ccTGA with a systemic RV and the complete AVB requiring conventional pacemaker therapy, which in the presence of ventricular dysfunction and conduction disease may further compromise cardiac performance, a biventricular pacemaker (CRT-P) was implanted. After cannulation of the coronary sinus (CS), an occluded CS venogram was obtained (Fig. 2) with a balloon tip catheter. A specially designed over the wire CS pacing lead (Attain OTW 4193, Medtronic, USA) was then advanced over the wire into the posterolateral cardiac vein (PLCV). A Medtronic 5076 screw-in lead was placed in pulmonic left ventricle (LV) septum. Subsequently, a bipolar active fixation lead (Capsure 5076, Medtronic) was positioned at the right atrium appendage. No phrenic nerve stimulation at maximal output was noted. The pacing and measurement of bioelectrical parameters at all the leads appeared satisfactory. These three leads were attached to a Medtronic pacemaker Syncra CRT C2TR01 (Fig. 3 Panel A and B).

Immediately after the introduction of CRT pacing, the sinus rhythm was reestablished and atrio-ventricular synchronization was achieved. The postoperative ECG showed an atrial and ventricular paced rhythm with a negative paced QRS configuration in DI lead and QRS duration of 130 ms (Fig. 4). The atrio-ventricular (AV) delays were programed as follows: AV delay sensed at 110 ms, AV delay paced at 130 ms, without VV delay.

Subsequently, the patient was discharged from hospital and put on captopril, carvedilol, spironolactone and aspirin. The follow-up was carried out every six months following the procedure. Postoperatively, the patient achieved total relief from her dizzy spells and her exercise tolerance improved from New York Heart Association functional Class III to II with a significant increase in her systemic right ventricle ejection fraction (RVEF = 45%) evaluated six months later. During follow-up, both atrial and ventricular lead parameters were all within satisfactory ranges and no arrhythmias were observed.

Within a 48 months follow-up, the patient achieved successfully two pregnancies after resynchronization therapy. The mode of delivery was vaginal. In the case of the first pregnancy, she developed active labor at 37 weeks, and had a spontaneous vaginal delivery of a small for gestational age infant with a congenital heart disease. A postnatal diagnosis of an aortopulmonary window was made in the offspring who died 3 days after surgery. For the

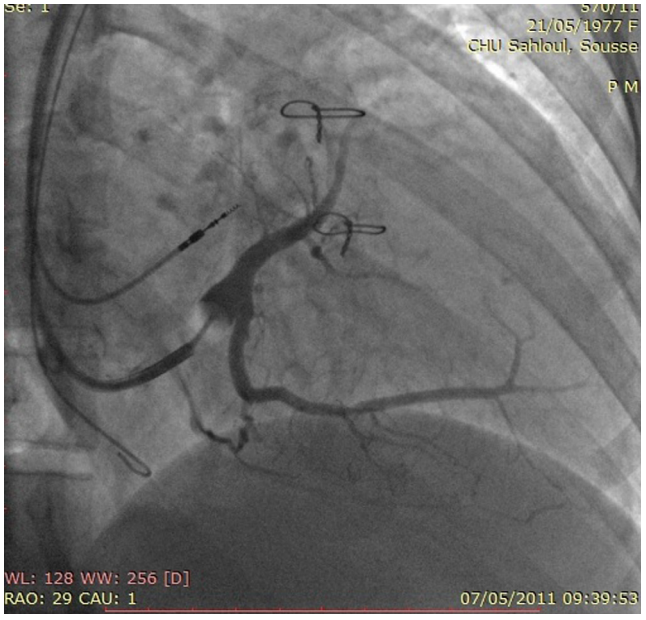

Fig. 2. Coronary sinus (CS) angiogram showing the CS tributaries in right anterior oblique view 30 (RAO 30°).

second pregnancy after resynchronization therapy, she tolerated labor and the childbirth occurred without any complication. The second infant had no evidence of structural heart disease on fetal or postnatal echocardiography or physical examination.

The RV ejection fraction was stable after the two pregnancies and estimated to be 43%. She remained active and had NYHA class II symptoms on medical regimen.

3. Discussion

We described a case of cardiac resynchronization therapy in 33 years old women with congenitally corrected transposition of the great arteries with complete AVB and systemic ventricular dysfunction who achieved six successful pregnancies and vaginal deliveries, four before and two after cardiac resynchronization therapy.

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