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## **The effect of comprehensive management of heart failure in adult with systemic right ventricle**

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Patients with systemic right ventricle (SRV) represent a significant proportion of adults with congenital heart defects (CHD) [1]. Over time, most of them show various degree of heart failure (HF) symptoms [1–5]. The lack of clear guidelines based on hard evidences and the increasing number of adult patients with CHD makes the treatment of SRV failure one of today's greatest challenges.

We present a case of 40 year old woman with congenital corrected transposition of great arteries, double-outlet right ventricle, ventricular septal defect, dextrocardia, after surgical closure of septal defect at the age of five. She was admitted due to symptoms of HF in New York Heart Association class III — exertional dyspnoea, many episodes of presyncope. From her pediatric period until now, she has been under the care of regional cardiology clinic, without any pharmacotherapy.

Transthoracic echocardiography (TTE) revealed significant enlarged SRV with poor systolic function (11 mm tricuspid annular plane systolic excursion [TAPSE] and 6.4 cm/s if longitudinal velocity of the tricuspid annulus [S'] evaluated by tissue Doppler),

moderate/severe systemic and moderate mitral valves regurgitations (Figure 1A). The ejection fraction of SRV in cardiac magnetic resonance imaging was 18%. The N-terminal pro-B-type natriuretic peptide was 5856 pg/ml. The initial maximal oxygen consumption ( $VO_2\text{max}$ ) was 16.6 ml/min/kg. However, exercise test was complicated with significant drop in blood pressure and prodromal symptoms of syncope in recovery. In Holter electrocardiography: significant ventricular arrhythmia with 14 episodes of nonsustained ventricular tachycardia (nsVT), right bundle branch block, QRS widening up to 200 ms, first-degree AV block (Figure 1B). The patient received pharmacotherapy recommended in classical HF: sotalol  $2 \times 40$  mg, dapagliflozin 10 mg, sacubitril/valsartan 24/26 mg, spironolactone 25 mg.

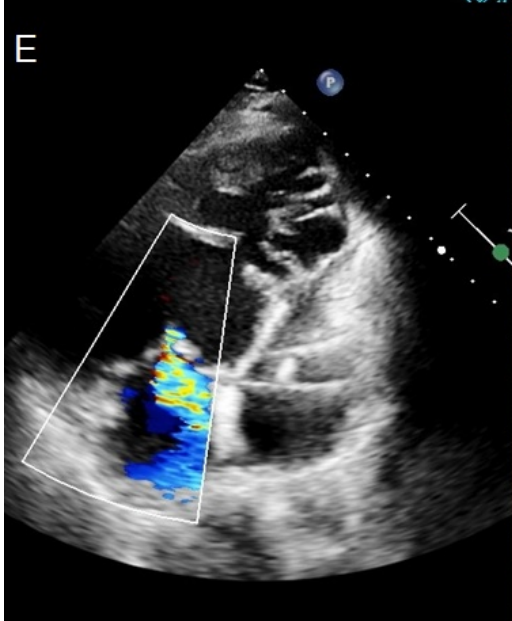
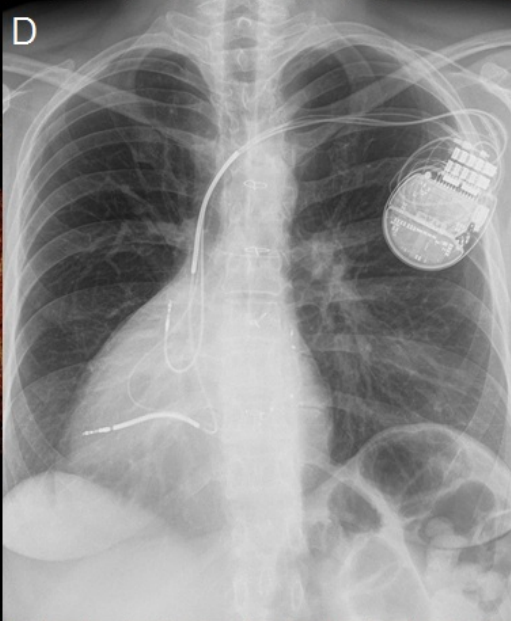
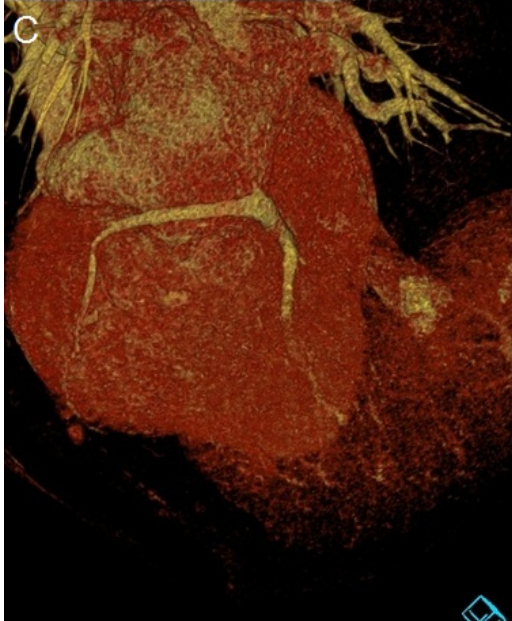
Moreover, she was qualified for urgent cardiac resynchronization therapy (CRT-D) implantation with computed tomography (CT) guidance (Figure 1C). Chest X-ray in Figure 1D shows final position of leads. After 4 months, control of CRT-D revealed a high latency with approximately 300 msec between pacing peak and left ventricular (LV) response. After exclusion of electrode dislocation, optimization of CRT-D system settings was performed with transthoracic echocardiography assistance from the M3-SVC vector biventricular pacing (LV to SRV) with LV preexcitation of 80 ms. Sotalol was changed to bisoprolol 2.5 mg.

At follow-up after 12 months, the patient reported significant improvement in exercise tolerance and New York Heart Association class II symptoms.  $VO_2\text{max}$  increased to 22.3 ml/kg/min without any worrisome effort-related symptoms. The echocardiographic picture of SRV was stable with ejection fraction estimated at around 20% (2D SRV volume quantification and visual assessment by a very experienced echocardiography specialist in modified SRV focused view), improved TAPSE to 12 mm and S' to 7.5 cm/s, moderate/severe systemic atrioventricular valve regurgitation (Figure 1E). However, N-terminal pro-B-type natriuretic peptide decreased to 3832 pg/ml. In Holter electrocardiography average heart rate was 64/min with stimulation of 97% with 1845 ectopic ventricular beats but without nonsustained ventricular tachycardia episodes (Figure 1F). Due to the tendency to hypotension, the doses of the drugs were not escalated.

Our case confirms the efficacy and safety of comprehensive management of SRV failure. However, this subject is still debatable [1–5]. A recent study based on the German National Register for Congenital Heart Disease revealed that in population with SRV polypharmacy with cardiovascular drugs was rare (4.5%) and 38.5% of the patients did not take any medication [5]. The impact of comprehensive management including sacubitril/valsartan and flozins in the treatment of SRV failure should be further investigated.

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**Figure 1.** **A.** Apical four-chamber view in initial ECHO. Enlarged systemic right ventricle (SRV). **B.** Image from Holter electrocardiography (ECG) recording before the introduced treatment. **C.** 3D reconstruction from a computed tomography angio performed before cardiac resynchronization therapy (CRT-D) device implantation procedure with coronary sinus imaging. **D.** Chest X-ray after implantation of CRT-D showing final position of leads. **E.** Apical four-chamber view in echocardiography conducted after comprehensive treatment. The stable picture of SRV with ejection fraction estimated at around 20% and moderate/severe systemic atrioventricular valve regurgitation. **F.** Holter ECG monitoring record after successful CRT-D implantation with apparent shortening of QRS complexes