The effect of comprehensive management of heart failure in an adult with a systemic right ventricle

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

We present a case of a 40-year-old woman with corrected congenital transposition of great arteries, double-outlet right ventricle, ventricular septal defect, and dextrocardia, after surgical closure of septal defect at the age of five. She was admitted for symptoms of HF in New York Heart Association (NYHC) class III — exertional dyspnea and multiple episodes of presyncope. From her pediatric care period to her current admission, she had been under the care of a regional cardiology center, without any pharmacotherapy.

Transthoracic echocardiography (TTE) revealed a significantly enlarged SRV with poor systolic function (11 mm tricuspid annular plane systolic excursion [TAPSE] and 6.4 cm/s when the longitudinal velocity of the tricuspid annulus [S'] was evaluated by tissue Doppler), moderate/severe systemic and moderate mitral valves regurgitations (Figure 1A). SRV ejection fraction 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_2max) was 16.6 ml/min/kg. However, an exercise test was complicated by a significant drop in blood pressure and prodromal symptoms of syncope in recovery. Holter electrocardiography showed significant ventricular arrhythmia with 14 episodes of nonsustained ventricular tachycardia (nsVT), right bundle branch block, QRS widening up to 200 ms, and first-degree AV block (Figure 1B). The patient received pharmacotherapy recommended in classical HF: sotalol 2 × 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). The chest X-ray in Figure 1D shows the final position of the leads. After 4 months, CRT-D control revealed high latency with approximately 300 msec between the pacing peak and the 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 NYHA class II symptoms. VO_2max 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 a modified SRV-focused view), improved TAPSE (12 mm), and S' (7.5 cm/s), moderate/severe systemic atrioventricular valve regurgitation (Figure 1E). However, the N-terminal pro B-type natriuretic peptide level decreased to 3832 pg/ml. On Holter electrocardiography, the average heart rate was 64/min with stimulation of 97% with

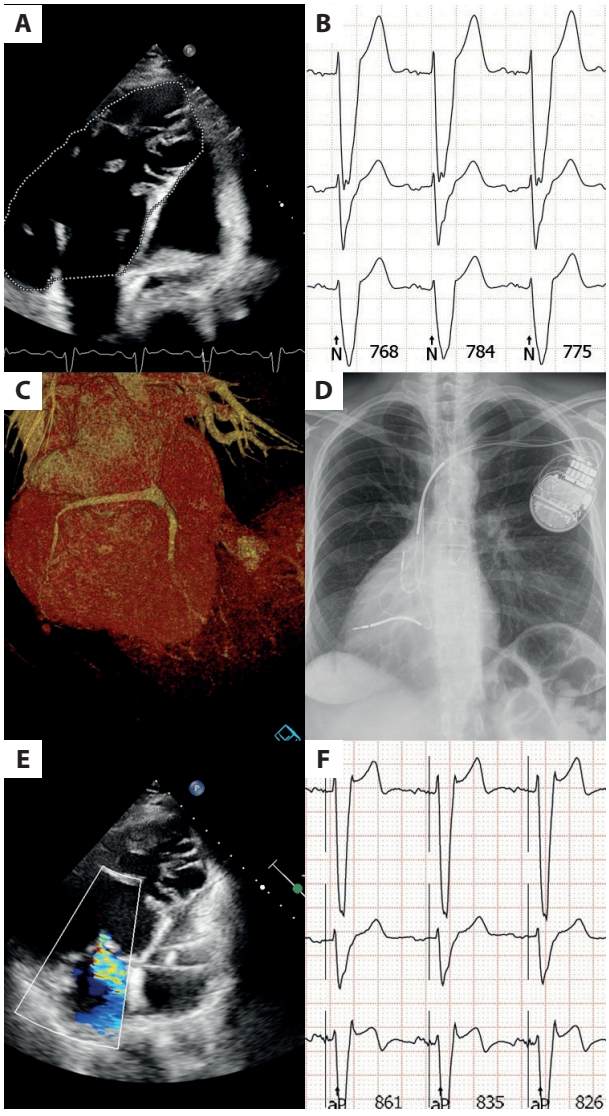

Figure 1. A. Apical four-chamber view on initial echocardiography. Enlarged systemic right ventricle (SRV). **B.** Image from Holter electrocardiography (ECG) recording before treatment. **C.** 3D reconstruction from computed tomography angiography performed before the procedure of cardiac resynchronization therapy (CRT-D) device implantation, with coronary sinus imaging. **D.** Chest X-ray after implantation of CRT-D showing the final position of the leads. **E.** Apical four-chamber view on 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 recording after successful CRT-D implantation with apparent shortening of QRS complexes

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 the SRV population cardiovascular drug polypharmacy was rare (4.5%), and 38.5% of patients did not take any medication [5]. The impact of comprehensive management including sacubitril/valsartan and flozins in SRV failure treatment should be further investigated.

Article information

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REFERENCES

1. Brida M, Diller GP, Gatzoulis MA. Systemic right ventricle in adults with congenital heart disease: anatomic and phenotypic spectrum and current approach to management. Circulation. 2018; 137(5): 508–518, doi: 10.1161/CIRCULATIONAHA.117.031544, indexed in Pubmed: 29378757.
2. Lluri G, Aboulhosn J. The systemic right ventricle in adult congenital heart disease: why is it still such a challenge and is there any hope on the horizon? Curr Opin Cardiol. 2022; 37(1): 123–129, doi: 10.1097/HCO.0000000000000933, indexed in Pubmed: 34857720.
3. Nederend M, Kiès P, Regeer M, et al. Tolerability and beneficial effects of sacubitril/valsartan on systemic right ventricular failure. Heart. 2023; 109(20): 1525–1532, doi: 10.1136/heartjnl-2022-322332, indexed in Pubmed: 37169551.
4. Neijenhuis RML, Nederend M, Jongbloed MRM, et al. The potential of sodium-glucose cotransporter 2 inhibitors for the treatment of systemic right ventricular failure in adults with congenital heart disease. Front Cardiovasc Med. 2023; 10: 1093201, doi: 10.3389/fcvm.2023.1093201, indexed in Pubmed: 37435053.
5. Lebherz C, Gerhardus M, Lammers AE, et al. Late outcome, therapy, and systemic ventricular function in patients with a systemic right ventricle: data of the German National Register for Congenital Heart Defects. Cardiol Young. 2022; 32(8): 1235–1245, doi: 10.1017/S1047951121003954, indexed in Pubmed: 34658317.