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Long-term outcome of patients with congenital heart disease undergoing cardiac resynchronization therapy

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Objectives: Cardiac resynchronization therapy (CRT) is rarely used in patients with congenital heart disease (CHD) and follow-up in available studies is short. We sought to evaluate long-term impact of CRT in patients with CHD.
Methods: Thirty consecutive patients with structural CHD (N=28) or congenital atrioventricular block (N=2) aged median 12.9 (IQR 6.5-18.2) years at CRT-P implantation were followed for median 9.0 (IQR 4.5-11.4) years on CRT. CRT was performed for systemic left ventricular (LV, N=12) and right (RV) or single (SV) ventricular (N=18) failure and was associated with additional cardiac surgery in 13 patients. CRT response was defined as an increase in systemic ventricular ejection fraction or fractional area of change (FAC) by >10 points and improved or unchanged NYHA class at the end of follow-up. Actuarial survival probability was calculated for 5 and 10 years after CRT implantation.
Results: Freedom from cardiovascular death or heart failure hospitalization was 92.0 and 82.3 %, respectively. No patient underwent heart transplant. Surgical revision of the pacing system was performed in 3 patients and pacing system extraction due to infection in 1 patient. CRT therapy was terminated in other 5 patients due lead dysfunction (exit block). Freedom from CRT complications leading to surgical system revision (elective generator replacement excluded) or therapy termination was 81.4 and 70.1 % at 5 and 10 years, respectively. Overall probability of an uneventful therapy continuation was 74.6 and 56.3 %, respectively. Upgrade to CRT-D was performed in 1 patient. Ejection fraction or FAC changed from median 29.5 % (IQR 22.3-35.0) before CRT to median 40.3 % (IQR 32.0-54.0; $p<0.001$) at the end of follow-up. NYHA class improved from median 2.0 to 1.5 ($p=0.002$). CRT response tended to be more frequent in systemic LV (9/12 patients) than RV or SV (6/18 patients, $p=0.060$).
Conclusions: Long-term CRT in patients with CHD is associated with acceptable survival and sustained improvement of systemic ventricular function in 50 % of patients. Probability of device complications necessitating surgical revision or therapy termination is however high.