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

Kubuš P. (1), Rubáčková Popelová J. (2), Kovanda J. (1), Sedláček K. (3), Janoušek J. (1)
Children’s Heart Centre, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital, Prague, Czech Republic (1); Department of Cardiac Surgery, Na Homolce Hospital, Prague, Czech Republic (2); Cardiology Department, Institute for Clinical and Experimental Medicine, Prague, Czech Republic (3)

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.