

Cardiac resynchronization therapy for treatment of chronic pulmonary right ventricular dysfunction in congenital heart disease

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Introduction: Chronic pulmonary right ventricular (RV) dysfunction is common in congenital heart disease (CHD). We sought to evaluate mid-term results of permanent RV cardiac resynchronization therapy (RV-CRT) in this setting.

Methods: Six patients with repaired tetralogy of Fallot (3), pulmonary atresia (1), absent pulmonary valve syndrome (1) and Ebstein's anomaly (1) having complete right bundle branch block and aged median 13.6 (1.0–19.2) years underwent RV-CRT for symptomatic chronic RV dysfunction/failure. Implantation procedures were associated with other cardiac surgery aimed to improve hemodynamic residua in 3/6 patients. RV-CRT was accomplished by atrial-synchronized RV free wall pacing at sites of late RV activation (q-RV interval = mean 83 ± 7 % of QRS duration) in maximal fusion with intrinsic ventricular activation as checked by regular 24-hour ECG. Patients were followed-up prospectively for median 14.3 (1.3-46.4) months and assessed by conventional echocardiography and speckle tracking imaging.

Results: All patients survived and improved clinically as well as in terms of RV function and synchrony (Table). RV-CRT had to be terminated after 3.2 years in one patient because of exit block on the RV lead. None of the patients experienced worsening of left ventricular function due to RV pacing.

	QRS duration [ms]	NYHA class	NTproB NP [ng/l]	RV FAC [%]	RV max. +dP/dt [mmHg/s]	RV MPI	Septal flash [N]	RV septal to free wall mechanical delay [ms]
Before	158 (29)	2.3 (0.8)	842 (756)	18 (9)	316 (153)	0.48 (0.44)	5/6	149 (72)
After	113 (20)	1.3 (0.4)	229 (196)	34 (4)	444 (161)	0.26 (0.10)	0/6	23 (70)
P	=0.002	=0.048	=0.118	=0.006	=0.051	=0.247	=0.015	=0.044

Mean (SD). FAC= fractional area of change; MPI=myocardial performance index

Conclusions: Permanent pulmonary RV-CRT alone or in association with redo cardiac surgery carried sustained clinical benefit and improvement in RV function and synchrony over mid-term follow-up in patients with CHD and RV dysfunction associated with electromechanical dyssynchrony. RV-CRT may be a useful adjunct to long-term management of RV dysfunction and failure in selected patients.