Management and Outcome of Patients with Double-Chambered Right Ventricle – Experience from Two Tertiary Adult Congenital Heart Disease Centers

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**Background:** Double-Chambered Right Ventricle (DCRV) is a rare form of congenital heart disease, in which the RV is progressively separated into a high- and low-pressure chamber. So far, there is no consensus on the optimal management of DCRV in adult patients. Here we report the combined experience with DCRV patients at two tertiary Adult Congenital Heart Disease Centers.

**Methods and Results:** All patients with DCRV under follow-up at the centers were identified from computerised databases. A retrospective review of medical records was carried out on all 53 identified patients (34 female, age 40.6 ± 15.0 at the time of data analysis, range 17 to 69 years). Almost all patients (96%) had ventricular septal defects (VSD) as an underlying diagnosis; predominantly in the perimembranous portion of the septum (80%). Seven patients had undergone VSD closure during childhood but continued to have a restrictive VSD. Eight patients were completely asymptomatic and were managed conservatively so far (median age 26 years, 34.4 ± 26.2, 19 to 63) with no fatalities. The remaining patients developed symptoms at a median age of 26 years (shortly after birth to 68 years), most commonly shortness of breath and reduced exercise capacity (56%), palpitations (13%) and chest pain (13%). Investigation by echocardiography revealed a mean intra-chamber gradient of 69.4 ± 30.0 mmHg. Surgical relief of the right ventricular obstruction was undertaken in 36 patients (68%, median age at operation 26.7 years, 28.3 ± 20.3, 14 months to 67 years.). There was no early or late operative mortality, no patient required reoperation for DCRV during follow-up and 86% of operated patients showed no residual intra-ventricular gradient postoperatively. Symptoms improved significantly after surgery with only 8 patients remaining in NYHA class 2 (mean follow-up time 10.5 ± 9.4 years.).

**Conclusions:** Contemporary DCRV patients, managed at tertiary congenital centers, have good survival prospects and low long-term morbidity. Cardiac surgery is inherently low risk, associated with good long-term haemodynamic and functional results and a proactive treatment approach therefore appears warranted.