

The right ventricular growth after decompression in patients with critical right ventricular outflow tract obstruction

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Objectives:To evaluate the right ventricular growth of patients in whom the initial management was right ventricle decompression. Patients with RV dependent coronary circulation were excluded from this study.

Methods:Between May 1994 to May 2010, 48 patients (27 M, 21F) with critical pulmonary stenosis (CPS) or pulmonary atresia with intact ventricular septum (PAIVS), (19 PAIVS, 29 CPS) underwent RV decompression. The mean age at first intervention was $4\pm 4,3$ days (range 1-19 days). Mean weight was $3,05\pm 0,5$ kg (range 1,9-4,6kg). Right ventricular development was assessed using tricuspid valve dimensions (TV) and RV length. TV was measured retrospectively on the cross-sectional echocardiograms performed before the procedure and during follow-up. Z score were used to standardize tricuspid valve dimensions with body size. The RV length was assessed from the tricuspid valve annulus to the RV apex at ventricular end diastole. At the latest review, we have evaluated 32 patients (17 CPS, 15 PAIVS). TV valve diameter at birth in these 32 patients ranged from 8 to 18 mm ($11,5\pm 2,68$), Z score of the TV valve ranged from -2,3 to 2,87 ($-0,08\pm 1,3$).

Results:During the follow-up, 29 patients out of 32 have a biventricular circulation, 2 patients have undergone 1 and $\frac{1}{2}$ ventricle repair, and 1 patient has undergone Fontan operation. Twelve of 32 patients (Group 1) required additional transcatheter or surgical procedures to augment the pulmonary blood flow. For group 1, TV diameter at birth ranged from 9 to 18 mm ($12,3\pm 3,3$). Z value ranged from -1,9 to 2,87 ($0,3\pm 1,5$). During the follow-up, TV diameter ranged from 19 to 31 ($23,5\pm 4$). Z score ranged from -6,1 to -0,48 ($-3,4\pm 1,6$). 29 of 32 patients (group 2) did not require additional procedures. For group 2, TV diameter at birth ranged from 8 to 14 mm ($11,1\pm 2,1$). Z score ranged from -2,3 to 1 ($-0,3\pm 1$). During the follow-up TV diameter ranged from 20 to 37 mm ($27,3\pm 4,2$). Z score ranged from -3,5 to 2,02 ($-1,7\pm 1,5$). RV length for group 1 ranged from 21 to 52 mm ($37,6\pm 10,8$) and for group 2 it ranged from 26 to 52 mm ($39,6\pm 8,5$).

Conclusion:Our data demonstrate that even in cases in which biventricular circulation can be achieved the growth of the right ventricle is not normal. It is important to investigate other factors such as genetic determinants that could influence right ventricular development.