Double Outlet Right Ventricle with Non-Committed Ventricular Septal Defect

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Objective: The management of Double Outlet Right Ventricle (DORV) associated with anatomically non-committed Ventricular Septal Defect (NCVSD) constitutes a surgical challenge. The limits for, and the specific outcomes after anatomical versus univentricular repair still remain to be established.

Methods: Between 1993 and 2011, 35 consecutive patients presenting with DORV/NC-VSD and 2 adequately sized ventricles were included into the study at two centers forming the National Referral Center. The selection criteria included the absence of outflow tract VSD: 21 inlet (4 complete atrio-ventricular septal defect (AVSD)), 9 muscular and 5 perimembranous. RVOTO was present in 18/35 (51%). Twenty patients had undergone 25 initial palliative procedures.

Results: Anatomical repair by means of intraventricular baffle construction was performed in 23 (Group I) at a median age of 10.5 months. VSD was surgically enlarged in 11 (48%). An associated RVOT reconstruction was required in 11 and Arterial Switch Operation (ASO) was done in 5. The remaining 12 patients underwent univentricular palliative repair (Group II). There were 4 hospital deaths (11.4%): 3 in Group I and one in Group II (p=.06). 8/20 survivors of group I patients underwent 13 reoperations after a median delay of 24 months, with subaortic stenosis being the main cause for reoperation (6/8). There was one late death in group 2. At last visit, all survivors were in NYHA class I-II. Ten years actuarial survival rate and freedom from reoperation were respectively 74.7 ± 5% and 58 ± 5 % in Group I, and, 80 ± 7% and 71 ± 7% in Group II. Univariate analysis showed that AVSD and/or isolated mitral cleft were associated with death (p=.04) and need for reoperation (p=.038).

Conclusions: Despite the need for complex procedure and the high incidence of reoperation for subaortic obstruction, our results suggested the long-term advantages of anatomical repair in DORV with NCVSD. The presence of associated AVSD and/or isolated mitral cleft was the only risk factors for mortality and reoperation.