Biventricular Repair of Complex Heterotaxy-Associated Congenital Heart Disease

OBJECTIVE: Heterotaxy-associated complex congenital heart disease (CHD) presents unique challenges to establishing biventricular circulation. We aimed to describe trends and contemporary outcomes in heterotaxy patients in whom biventricular repair (BVR) is attempted.

METHODS: All patients with heterotaxy and complex CHD who underwent surgical palliation at a quaternary pediatric cardiac center from 2006-2022 were stratified by single ventricle palliation (SVP) vs. 1.5-ventricle or BVR.

RESULTS: Amongst 210 heterotaxy patients with complex CHD, 180 (86%) underwent SVP, and BVR was attempted in 30 (14%). Atrioventricular canal (AVC) defect was present in 121 (67%) SVP patients (85 right dominant, 26 left, 10 balanced) vs. 14 (47%, p=0.03) BVR patients (7 right dominant, 1 left, 6 balanced). 136 (76%) in SVP had a dominant right ventricle (RV) vs. 11 (37%, p<0.001) BVR. There was a strong era effect on percentage of BVR, consistent with evolution in seeking biventricular circulation for these complex defects – 3/63 (5%) BVR in the 1st tertile vs. 10/79 (13%) in 2nd and 17/68 (25%) in 3rd (p=0.02). Median follow-up was to 6 (2.4-8) years of age. Of the 180 SVP patients, 74 (41%) have reached Fontan, 6 (3%) and 31 (17%) await Glenn and Fontan, respectively. In BVR cohort, 18 (60%) have reached biventricular circulation, 2 (7%) 1.5-ventricle, and 2 (7%) await BVR. There were 62 (30%) operative mortalities during any surgery, 55 (31%) SVP and 7 (23%, p=0.7) BVR. 35 (56%) patients who died had AVC, 30 (55%) SVP and 5 (71%) BVR. Presence of AVC was an independent predictor of mortality in the BVR group only (p=0.04). TAPVR was present in 31 (50%) - 29 (53%) SVP and 2 (29%) BVR (p=0.2). 48 (77%) patients who died had a primary RV, 43 (78%) SVP and 5 (71%) BVR (p=0.7). An additional 14 (8%) SVP patients died interstage and 12 (7%) post-Fontan. In the BVR group, 1 (3%) patient died awaiting BVR after ventricular recruitment, and there were no deaths in patients who reached 2- or 1.5-ventricle physiology.

CONCLUSIONS: Outcomes continue to be sub-optimal in patients with heterotaxy-associated complex CHD. Although more of these patients are being considered for BVR, this proportion remains small. The presence of AVC is associated with increased mortality in heterotaxy patients who undergo BVR.

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