Longterm Follow-up After Biventricular Repair of the Hypoplastic Left Heart Complex

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Introduction: In hypoplastic left heart complex (HLHC) patients univentricular palliation can be avoided with more favorable results from biventricular repair. However, studies on patient outcome after biventricular repair are limited and show divergent results. We retrospectively characterized patients with HLHC after biventricular repair, analyzed mid-and longterm outcome and assessed left heart structures growth.

Methods: Patients with HLHC after biventricular repair between 2004 and 2018 were retrospectively reviewed. HLHC was defined as an aortic valve (AoV) or mitral valve (MV) annulus Z-score of less than -2 and a coarctation or aortic arch hypoplasia. Clinical outcome was analyzed and MV and AoV annulus, left ventricular (LV) length and LV internal diastolic diameter (LVIDd) were measured before surgery and 6 months and 1, 3, 5 and 10 years after biventricular repair.

Results: In 30 patients, median age at surgery was 10 days (IQR, 7.75 to 13.25). All patients survived. Fourteen patients (46.7 %) required a surgical or catheter based reintervention. Median follow-up was 6.19 years (IQR, 4.12 to 10.16). Mean preoperative Z-scores ± SD (range) were calculated for MV (-2.82 ± 0.96 (-5.17 to -1.10)), AoV (-2.29 ± 1.22 (-4.85 to -0.25)), LV length (-2.09 ± 1.05 (-4.77 to -0.23)) and LVIDd (-2.24 ± 1.61 (-6.45 to 0)). Paired T test showed that most of the catch up growth was seen in the first 6 months after repair, as indicated by the significant difference between mean preoperative and 6 months follow up Z-scores: MV mean difference 1.24, 95% CI: 0.86 to 1.61, AoV mean difference 1.97, 95% CI 1.46 to 2.48, LV length mean difference 1.60, 95% CI 1.12 to 2.09 and LVIDd mean difference 2.70, 95% CI 2.07 to 3.33. At 10 year follow-up all dimensions normalized, with limited growth of the MV (mean Z-score ± SD is -1.41 ± 0.64).

Conclusions: Biventricular repair in HLHC patients can be performed with good results, although almost half of the patients required a reintervention. Growth of left sided cardiac structures is most important in the first 6 months after repair with Z-scores almost normalized at 10 years follow up.