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Experience of cardiovascular surgery for congenital heart disease associated with trisomy 13 &18

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Objectives: this study was to examine a single institutional experience of patients with trisomy 13 and trisomy 18 in the setting of comorbid congenital heart disease (CHD) and present the outcomes of surgical intervention.

Methods: Patients with CHD and trisomy 13 or 18 presenting to our institution from 2009 through 2018 were retrospectively reviewed. Ten consecutive trisomy 18 patients and three consecutive trisomy 13 patients (13 patients in total) with comorbid CHD underwent surgical intervention. These 13 patients had ages ranging from 3-389 days (median 45 days), birth weight of 1378–3208 g (median 1972g), and length of gestation of 34–41 weeks (median 37). Palliative operations performed in 12 patients of ventricular septal defect (VSD), including pulmonary artery banding in 9, bilateral pulmonary artery banding for the treatment of HLHS variant in 1, and systemic-to-pulmonary shunt for the treatment of TOF/pulmonary atresia in 2 patients. Concomitant procedures included patent ductus arteriosus closure in 8 patients and a lung biopsy in 5 patients and were performed at a median age of 45 days (3-389 days) and a median weight of 2.15 kg (1.3-5.8 kg). Two patients underwent subsequent second-stage intracardiac repair electively. Primary intracardiac repair consisted of VSD closure and patent ductus arteriosus closure in one patient.

Results: The hospital mortality was 15% (n = 2), and 11 patients (85%) were discharged home with improved symptoms. The survival from surgical intervention ranged from 6-115 months (median 31 months). Two patients experienced late death due to non-cardiac events (respiratory failure and hepatoblastoma in one patient each).

Conclusions: Our data suggest that cardiac surgery may improve the survival in select patients with trisomy 13 and 18. However, the indication for surgery should be carefully considered on a case-by-case basis, as there remains a risk of late death even after surgery. Even if the patient can be discharged from the hospital after surgery, the family may often be overwhelmed by the care, such as respiratory support, tube feeding and have severe mental retardation for multiple congenital anomalies.