Prognostic factors for early outcomes in patients with total anomalous pulmonary venous return


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The aim of this study was to assess postoperative outcomes and pronostic factors of children with total anomalous pulmonary venous return (TAPVR)

Methods: all cases diagnosed with TAPVR from 1973 to 2014 were included. Clinical, echocardiographic, surgical data were collected and analyzed. Patients were divided into 4 groups according to decades of follow-up and comparisons were made between groups to assess changes over time.

Results: 180 patients were included= 78 supracardiac 43%, 48 intracardiac 27%, 35 infracardiac 19% and 19 mixed 11%. Pulmonary venous return was obstructed in 45%, more frequently in infracardiac type (69%). Mean age at diagnosis was overall 88 days, lower in infracardiac group (11 days). Age at surgery was 107 days, decreased from 111 days to 18 days over time. Heart failure was the leading symptom at diagnosis (111/180 cases) and 34% of the cases needed mechanical respiratory support before surgery. Associated lesions were present in 33 cases=18.3% (the most frequent was VSD : 15/33). Mean weight at surgery was 4.1kg (min 1.8kg). Delayed chest closure was observed in 58% of infracardiac cases (overall 18.3%). Mean postoperative duration of MVS, CICU stay and hospital stay were respectively 7.4, 10.1 and 18.4 days. Postoperative in-hospital mortality decreased over decades from 42% before 1980 to 7.4% currently. Decade, duration of bypass, body weight, suprasystemic pulmonary pressure and preoperative acidosis were risk factors for in-hospital death. Arrhythmias and pulmonary hypertension acute crisis occurred in 11% and 10.7% during the early post-surgical course. Mean pulmonary pressure decreased from 68mmHg preoperatively to 40mmHg within the 1st week after surgery. Inhaled NO was used since 1992 and had a significant beneficial impact on survival (20-year survival 85% versus 55%, p< 0.0001).

Conclusion: Overall prognosis of patients with TAPVR improved over decades and NO significantly contributed to improve survival. Patients with infracardiac TAPVR have more severe clinical presentation and worse early outcomes.