Long term outcome and survival of neonates with total anomalous pulmonary venous return

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The aim of this study was to assess long term survival of children with total anomalous pulmonary venous return (TAPVR).

Methods: Clinical, echocardiographic, and long term complications and reinterventions were assessed in all cases diagnosed with TAPVR from 1973 to 2014.

Results: 180 patients were diagnosed with TAPVR: 78 supracardiac 43%, 48 intracardiac 27%, 35 infracardiac 19% and 19 mixed 11%. Mean FU was 11 years. One hundred and forty two patients survived in-hospital stay (79%). The incidence of late complications was 19.6% (28 cases): 8 pulmonary veins stenosis, 5 residual ASD or partial APVR, 3 vena cava stenosis, 3 right to left atrial shunt, 3 neurological sequelae and 6 miscellaneous.

Duration of bypass, mixed pattern TAPVR and preoperative mechanical ventilatory support were significant risk factors for late complication respectively p= 0.01, p= 0.0023 and p= 0.09). Eight patients suffered from late pulmonary venous stenosis, of whom 7 died (6 in the early course after redo surgery and 1 before reoperation) and 6 were operated on before 2000.

Sixteen patients were reoperated on for pulmonary venous stenosis (7), residual ASD or partial PVR (5) and vena cava stenosis (4). Twelve late deaths occurred (6.7%), 5 from non cardiac causes and 7 due to pulmonary veins stenosis.

Survival was 80%, 75%, 70% and 65% at respectively 1 year, 10 years, 20 years and 40 years of FU. Survival was significantly lower in infracardiac type (p= 0.0017). Overall 131 patients survived of whom 84.7% are in NYHA class I with none ongoing cardiac medication. Pulmonary pressure levels range within normal value in 109 cases (83%), grade I PHT persists in 9 (7%).

Conclusion: Overall late outcomes of patients with TAPVR is favourable, and the infracardiac type is associated with the lowest survival rates. Pulmonary veins stenosis is a rare but lifethreatening late complication.