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Total Anomalous Pulmonary Venous Connection: Clinical Presentation and Long Term Outcome of 60 Patients at a Single Institution in 32 Years

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Background: Total anomalous pulmonary venous connection (TAPVC) is notoriously difficult to diagnose antenatally as well as posing diagnostic challenges postnatally in very sick neonates. We aimed to report our 32 year experience on the clinical spectrum and outcome of TAPVD in Wales.
Patients and Methods: We reviewed clinical records of 60 patients underwent TAPVC repair between 1980 and 2013. Fifty patients had isolated TAPVC (group I), and 10 had associated complex congenital heart disease (group II).
Results: The anomalous drainage was supra cardiac in 28 (46.6%), cardiac in 14 (23.3%), infracardiac in 14 (23.3%), and mixed in 4 patients (6.6%). Major associated cardiac anomalies were present in 10 patients (6 right atrial isomerism + CAVSD + DORV, 2 RAI+CAVSD, 1 DILV, and 1 HLHS). The antenatal detection rate was 11%. The median age at diagnosis was 11 days (1-270 days) in group I, and one day (1-11 days) in group II. Cyanosis (51%) and respiratory distress (35%) were the main symptoms. 19 patients with isolated TAPVC had severe pulmonary hypertension (38%) and the initial diagnosis was persistent pulmonary hypertension in 5 neonates. Severe PV obstruction was found in 42% of infracardiac type, and in 14% of supracardiac type. The median age at operation was 28.5 days (1 day-7 years). The median follow up was 11.3 years (11 months-35 years). PV obstruction developed in six patients, baffle obstructıon and narrowing of SVC occurred in 4. 18 patients (36%, 18/50) with total correction developed rhythm problems including right bundle block, low atrial rhythm, junctional rhythm (one requiring pacemaker) and atrial fibrillation. Six patients in group II (60%) and 2 in group I (4%) died in the follow up. The 1-year and 5-year survival rates were 96% in group I.
Conclusion: The antenatal detection of TAPVD is unacceptably low. Early recognition of TAPVD requires careful echocardiographic examination of newborns presenting with PPHN or severe respiratory symptoms. The outcome of isolated TAPVC is favorable, however when it is associated with major cardiac anomalies it has higher mortality and morbidity. The PV drainage site *per se* was not associated with adverse outcome.