Surgical repair of Total Anomalous Pulmonary Venous Connexion in neonates and infants: Immediate results and long-term follow-up.

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Introduction: The Total Anomalous Pulmonary Venous Connexion (TAPVC) represents 1 to 3% of congenital heart diseases. An early surgical repair is mandatory due to the severity of the hemodynamic impact. Objectives were to analyse a large population of patients operated for TAPVC with a special attention given to the post-operative mortality, risks factors, morbidity and long-term follow up.

Methods: a cohort of 92 neonates and infants referred in a single center between 2000 and 2014 were retrospectively reviewed. Median age at surgery was 9.5 days (range 1-143), 65.2% were neonates, 63% of patients were mechanically ventilated before surgery. TAPVC was supracardiac in 38%, intracardiac in 15.2%, infracardiac in 37% and mixed in 9.8%. Preoperative obstruction was found in 58.7% of cases, significantly associated with infracardiac connexion (OR = 5.74, 95% CL: 2.07-15.97, p <0.001). Patients with atrioventricular septal defect, heterotaxy with single ventricle, cor triatriatum or partial anomalous pulmonary venous connexion associated were excluded.

Results: Post-operative mortality was 4.3% (95% CL: 0-8.5). The median follow-up was 6.7 years (range: 0.3-13.4 years). The overall actuarial survival rate of operated patients was 86.5% (95% CL: 8.7-94.3) at one year, remaining stable beyond. Reoperations for pulmonary vein stenosis were required in 13 patients, leading to death in 23.1% (95% CL : 0.0-46.1). The first reoperation was performed after a median delay of 3.6 months (range: 0.4-127.1) from the surgical repair. In multivariate analysis, combination of heart defects associated with TAPVC (p<0.001), prematurity (p <0.001) and a persisting postoperative pulmonary hypertension (p=0.04) were found as risk factors for death. A postoperative portal venous thrombosis occurred in 4 patients after infracardiac TAPVC repair (n=34). All surviving patients were in NYHA class I after 5 years of follow up.

Conclusion: Surgical repair of TAPVC gives excellent immediate and long-term results. Mortality was limited to the first postoperative year. The main long term complication was the pulmonary vein obstruction that led to a high-risk reoperation. The occurrence of portal venous thrombosis after infracardiac TAPVC repair reached more than 10% of patients, justifying a systematic research.