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Experience with total cavopulmonary connection in children

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The objective of this study was to analyze a cohort of children with single ventricle heart after total cavopulmonary connection (TCPC) and assess the factors predictive of early and late outcome .

Methods: Patients who underwent TCPC at age < 18 years from 2000 to 2015 were retrospectively reviewed. Demographic data, cardiac disease, previous medical and surgical history, echocardiographic measurements, all events and outcomes were collected. Predictive factors for survival and events were assessed.

Results: 98 patients were included in the study, Age at TCPC was 7.9 ± 3.9 years (range 20 months to 17 years, media, 7.2). Underlying cardiac disease was: 26 tricuspid atresia, 13 double inlet ventricle, 11 pulmonary atresia, 11 unbalanced atrioventricular defect. 9 single ventricle with right outflow tract obstruction, 13 single ventricle with left outflow tract obstruction, 9 complex CHD, 5 hypoplastic left heart syndrome, and 1 Ebstein. Preoperative ventricular function was normal in 50%, mildly impaired in 50%. Age at TCPC was lower in the 2010-2015 period than before (mean 6.6 versus 8.9 years, $p=0.004$). Postoperative duration of drainage ranged 8 days to 9 months (median 10 days). Fenestration was performed in 7 cases (7.2%). One or more complications occurred in 27 cases (27.5%): arrhythmia in 6, heart failure in 2, PLE in 7 (7.2%). Two deaths occurred (2.2%) and 4 underwent HTX (4.2%), at a median time of 1.6 and 7 years after TCPC respectively. The incidence of PLE was 10.7% in cases operated before 2010 and 2.4% TCPC performed in 2010-2015 period. All deceased and transplanted cases had undergone TCPC before 2010. Age at TCPC did not differ between failed cases (i.e. death, HTX, PLE) and others. PLE was significantly correlated with death or transplantation (28.5% versus 4.4%, $p=0.01$). Overall survival was 97.9% at follow-up of 14 ± 7.3 years after TCPC (median 13.5 years).

Conclusion: TCPC survival rates in children is favourable and improved since 2010 while age at surgery was decreased. PLE is predictive of death or transplantation.