

The Outcomes of the Extracardiac Conduit Fontan Operation in Patients with Heterotaxy Syndrome: Experience in a Single Institute.

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Introduction

In patients with single ventricle physiology and heterotaxy syndrome, interim mortality and morbidity before completion of Fontan procedure have been known to be high. We sought to assess the outcomes after completion of Fontan procedure in heterotaxy patients compared with non-heterotaxy patients.

Methods

From December 1993 to July 2010, 235 patients have completed extracardiac conduit (ECC) Fontan palliation. We reviewed the results of these patients retrospectively and the data of subgroup having heterotaxy, 57 patients were compared with those who did not have heterotaxy syndrome.

Results

57 patients were comprised of 6 left isomerism and 51 right isomerism. Patients with heterotaxy syndrome had significantly higher number of total anomalous pulmonary venous return (59.6% versus 1.1%) and separate hepatic vein structure (29.8% versus 0.0%). Pulmonary vascular compliance (PA index/mean PAP) were higher in heterotaxy subgroup before Fontan operation. After ECC Fontan procedure, duration of pleural effusion, ICU stay and ventilator use were not significantly different from non-heterotaxy group. In long-term follow-up, grade of NYHA classification, degree of atrioventricular regurgitation, incidence of protein losing enteropathy and reintervention rate were not different in both groups.

Overall mortality rate were 10.1% during 10-year follow up period and there was no significant difference statistically between heterotaxy group and non-heterotaxy group in mortality. (5.1% versus 10.5%, $p=0.125$)

Conclusion

Heterotaxy syndrome was not increasing risk of mortality or morbidity once after completion of ECC Fontan operation. This study implies that heterotaxy syndrome has more anomalies in venous connection, however, its complications and prognosis were not different from other functional univentricular heart after Fontan palliation.