Late Sequels after Cavopulmonary Anastomosis in Complex Congenital Heart Disease: Thailand Experience

Noiming P., Tanpowpong P., Samankatiwat P., Limsuwan A.
Faculty of Medicine Ramathibodi hospital, Mahidol University, Bangkok, Thailand.

Introduction: The survival rate of children with congenital heart disease (CHD) has been improved. Cavopulmonary anastomosis (CPA) has become a standard procedure in patients with complex CHD particularly with single ventricle physiology. These children have been survived with the sequels impact of CPA. The purpose of study is to evaluate late complications in these patients.

Methods: Patients, underwent CPA at least 5 years and had been regularly followed at our institute, were recruited. Medical records had been retrospectively reviewed. Periodic cardiovascular investigations and additional hepatic evaluation had been obtained during Jan-Oct 2017.

Results: Thirty consecutive patients (20 male), were recruited with mean age of 14.8 years (7-33 years). Their median age at operation was 7.6 years old (3-23 years). Three of them (10%) underwent single-stage total CPA or Fontan versus 21 patients (70%) had staged Fontan operation. One fifth of the group (6 patients) underwent palliative inferior CPA or inferior-Glenn (inferior vena cava –to – pulmonary artery) because they presented beyond their infancy period and were not suitable for single-stage Fontan. Of all Fontan patients, 23/24 patients had extra cardiac conduit while 1 patient had lateral tunnel. Fenestration was performed in 23 patients. Median duration of follow up was 7.4 years (5-17 years). Majority of the patients (93.3%) were asymptomatic with NYHA class I or II. Late complications consist of dysrhythmia 16.7%, thromboembolism 16.7%, sinus node dysfunction required permanent pace maker 10%, and protein-losing enteropathy 6.7%. Almost all of the patients had increased hepatic stiffness detected by Fibroscan (which suggested liver fibrosis) while one of the patients had MRI confirmed hepatic nodule. In fenestrated Fontan group, 12 patients (52%) underwent device closure of fenestrations. There is no statistical significant difference of late complications between patency of fenestration versus device closer groups.

Conclusions: Long term follow up for group of Fontan and inferior-Glenn patients revealed satisfactory result with majority of the patients were in functional class I and II. However, there are major concerns regarding systemic venous hypertension and hepatic squeals. Further regularly systematic follow-up is required particularly evaluations for early hepatic fibrosis detection and management.