Current era outcomes of pulmonary atresia with ventricular septal defect : A single center cohort between 2005 and 2016

Kanthalas Lertsakulpiriya MD[1], Chodchanok Vijarnsorn MD[1], Prakul Chanthong MD[1], Paweena Chungsomprasong MD[1], Supaluck Kanjanauthai MD[1], Kritvikrom Durongpisitkul MD[1], Jarupim Soongswang MD[1], Thaworn Sabtaweesin MD[1], Somchai Sriyoscharti MD[1]

[1] Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Objectives: to evaluate survivals and prognosis of children who had diagnosis of pulmonary atresia with ventricular septal defect (PA/VSD) at Siriraj hospital in the decade.

Methods: A single-center retrospective study of children who had been referred for a management of PA/VSD with or without major aortopulmonary collateral arteries (MAPCAs) at Siriraj hospital between January 2005 and December 2016 were conducted. Patients who had single ventricle or previously undergone operation from other hospitals were excluded. Demographic data, operations and outcomes were reviewed. The survival analysis was performed at the end of 2018. Mortality risks were explored using multi-variate analysis.

Results: A total of 90 patients (44.4% male) were analyzed. A median age of referral and diagnosis at the center was 0.5 (0 – 13.8) years. 22 patients (24.4%) had other associated anomalies. Patients' pulmonary arteries (PAs) were categorized into 4 groups; 1) PA/VSD with confluent PAs (n=40), 2) PA/VSD, confluent PAs, MAPCAs (n=21), 3) PA/VSD, non-confluent PAs, MAPCAs (n=12), PA/VSD, small native PAs, MAPCAs (n=17). Of 88 patients who underwent operations, 32 patients; group 1 (n=15), group 2 (n=11), group 3 (n=4) and group 4 (n=2), had complete repair at 8.4 ± 4.6 years of age. During the follow-up [median time of 5.7 years (7 days - 13.6 years)], 17 patients (18.9%) deceased. Survival rates at 1, 5, and 10 years of age were 95.6%, 83.7% and 79.6%, respectively. Significant mortality risks were presence of associated anomalies and non-confluent PAs.

Conclusion: Children with PA/VSD have a moderately good survival in the current era. Despite multiple staged interventions, some patients can achieve operative correction. We report, herewith two predictors of death in this population.

Keywords: congenital heart disease; pulmonary atresia with ventricular septal defect; Survival rate; prognosis

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