

Advantages of hybrid management for pulmonary atresia, ventricular septal defect, major aorto-pulmonary collateral arteries and completely absent central pulmonary arteries

Hoashi T. (1), Kagisaki K. (1), Yazaki S. (1,2), Kitano M. (1), Kurosaki K. (1), Shiraishi I. (1), Yagihara T. (1,3), Ichikawa H. (1)

(1)National cerebral and cardiovascular center, Suita, Japan

(2)Sakakibara Heart Institute, Tokyo, Japan

(3)Rinku general medical center, Izumisano, Japan

Objectives: To review early and long-term clinical outcomes for pulmonary atresia, ventricular septal defect (PA/VSD), major aorto-pulmonary collateral arteries (MAPCAs) and completely absent central pulmonary arteries (cPAs)

Methods: Of all 120 patients with PA/VSD, MAPCAs who underwent surgical intervention between 1981 and 2011, 15 patients (12.5%) with completely absent cPAs were enrolled. Median age at initial surgery was 1.7 years-old (range, 0.1-17.6). Median number of unifocalized MAPCAs were 3.5 (2-6). Before 2005, various surgical approaches had been applied (Other group, n= 10), including staged unifocalizations via thoracotomy followed by RV-PA conduit placement and complete VSD closure in 8 patients, primary definitive repair via median sternotomy in 1, and the other in 1. Since 2003, hybrid management has been applied (Hybrid group, n= 5), where complete unifocalization and RV-PA conduit placement via median sternotomy as the 1st surgical palliation, then which was followed by VSD closure with one-way fenestrated patch and conduit replacement about 1 year later. For hybrid group, percutaneous trans-catheter balloon angioplasty (PTA) was aggressively performed after, and also before the 1st surgical palliation. After the definitive repair, stent implantation in addition to PTA was repeated for both group patients, if necessary. Follow-up was completed on all patients and median follow-up period was 7.8 years (0.3-21.7).

Results: Whereas all 5 patients in Hybrid group survived long after the definitive repair, 5 patients died before or early after the definitive repair in Other Group (p= 0.05). Uncontrollable bleeding complications were related to death in 4 of all 5 mortalities. Actuarial survival rates at 10 years after the initial surgery were 100% in Hybrid Group and 40.0% in Other group (log-rank, p=0.08). For 10 patients survived long after the definitive repair, catheter intervention for unifocalized MAPCAs were repeated in 6 patients and latest catheter examination at median duration of 4.8 years after the definitive repair showed that RV/LV pressure ratio was maintained from 0.60 (0.45-0.73) at early after the definitive repair to 0.57 (0.35-0.81) .

Conclusions: Although statistical significance was not proven, maintaining patency of MAPCAs by hybrid management can be expected to provide better early and long-term clinical outcomes.