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Preoperative dilatation of the right pulmonary artery is a risk factor for refractory respiratory complications late after the definitive repair of Tetralogy of Fallot with absent pulmonary valve

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Background: Refractory respiratory complication is not a rare condition late after definitive surgery of Tetralogy of Fallot with absent pulmonary valve (TOF/PVA). Here we evaluated perioperative risk factors for the post-operative respiratory complications in TOF/PVA patients.

Methods: Twenty-one patients with TOF/PVA who had underwent the definitive repair (1987-2012) were enrolled in this study. Plication of the pulmonary artery (PA) was performed in 19 patients. Cardiac catheterization was examined before surgery in 15 patients. We evaluated preoperative conditions of the patients as follows; age at surgery, sex, hemodynamic data in cardiac catheterization and those in echocardiography. We also evaluated the right and left pulmonary arterial area index (RPAI, LPAI) using cine-angiography; $\pi \times (\text{right or left pulmonary diameter} / 2)^2 / \text{body surface area}$. Clinically significant respiratory complications are defined as; 1) repeated admission due to respiratory infection and 2) mechanical ventilation over one month.

Result: One patient died during perioperative period and one died late after the operation. Overall survival rate was 86% in 20 years. Eight patients (38.1%) including two mortalities developed significant respiratory complication. One patient underwent tracheostomy and mechanical ventilation, seven patients needed unscheduled hospitalization due to repeated respiratory infections. These all eight patients had preoperative respiratory failure and required definitive surgery in the first year of life. Preoperative respiratory failure was a risk factor ($p=0.04$). Larger RPAI was also a significant risk factor of postoperative respiratory complication compare with those who had no respiratory complication (RPAI: 1371 ± 616 vs. 856 ± 298 ; $p=0.044$). Post-operative PAI did not show any significant difference in patients with or without post-operative respiratory complication ($p=0.657$). None of other perioperative factors including type of reconstruction of right ventricular outflow tract, preoperative LPAI and postoperative RPAI, data in echocardiography had a significant correlation with the postoperative respiratory complication.

Conclusions: Survival after the definitive repair for TOF/PVA was satisfactory. The postoperative respiratory complications were correlated to the preoperative respiratory failure and the dilatation of the right pulmonary artery. The post-operative outcomes could be improved by earlier surgical interventions before marked dilatation of the right pulmonary artery and severe respiratory disturbance develop.