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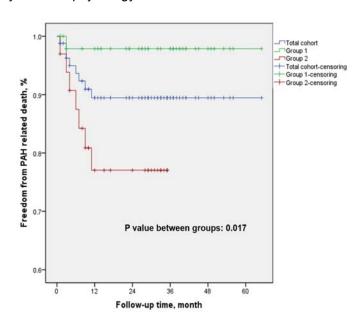
Pulmonary arterial hypertension related to complete transposition of the great arteries: outcomes after arterial switch

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Introduction: Transposed great arteries (TGA) is the most common complex congenital heart defect leading to pulmonary arterial hypertension (PAH). Clinical scenario of patients with TGA-PAH after arterial switch operation (ASO) remains largely unknown. This study aimed to investigate mid- to long-term outcomes focusing on pulmonary vascular physiology.

Methods: Consecutive patients from 2010 to 2014 were retrospectively included. The inclusive criteria included 1). diagnosed as TGA-unrestrictive VSD, 2). age > 6 months, 3). underwent ASO and 4). mPAP > 25 mmHg / PCWP < 15 mmHg. Patients co-present with collateral vessels, left ventricular outflow tract obstruction and cardiac positional abnormality were excluded.

Results: A total of 83 patients were included with an overall mortality of 13.3% (11/83). Fifty patients ≤ 1 years old were divided into group 1 and the others (33 patients) > 1 years old were in group 2. Eight deaths (72.7%) were related to PAH and all of them occurred within 1 year after ASO. The figure displays the probability freedom from



PAH related deaths, which was 93.6%, 89.5%, and 89.5% at 6 months, 1 year and 5 years, respectively. Only the mPAP immediately after ASO was identified as independent risk factor for mortality (OR=0.8, p=0.020) in multivariate analysis. Among the 33 patients with abnormal mPAP at discharge, the PAH related deaths were less in those with regular postoperative oral drug treating PAH (2/22 vs. 6/11, p=0.015).

Conclusions: Patients with an age <1 year can undergo ASO with favorable outcomes and have better survival rate than those older than 1 year of age. Deaths were associated with mPAP immediately after ASO. Regular oral drug is indicated in patients with abnormal postoperative mPAP.