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Pulmonary atresia and ventricular septal defect with major aorto-pulmonary collateral arteries

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Introduction: Pulmonary atresia (PA) with ventricular septal defect (VSD) is the ultimate form of tetralogy of Fallot, which comprises of 0.07 per 1000 live birth.

Case Description: A 3 years-old boy presented with recurrent dyspnea and fatigue since 6 months ago. He usually looks blue in the lips and fingers while crying; recurrent squat and failure to thrive also complained by his parent. He was born with spontaneous vaginal birth, weight of 3200 grams. He currently weighs 10.7 kg with stable vital signs. Resting oxygen saturation of 68-72% with clubbing finger was observed. Ejection systolic murmur grade II/6 at lower left sternal border and no continuous murmur was heard on auscultation. Blood and radiology workup indicated secondary polycythemia vera and increased bilateral perihilar markings. ECG showed RAD, P pulmonale and RVH with incomplete RBBB. Transthoracic echocardiography revealed bidirectional shunt of VSD, overriding aorta, absence flow from RV to PA with abundant of major aorto-pulmonary collateral arteries (MAPCAs). No left aortic arch CoA nor PDA was found. These findings suggested a PA-VSD with MAPCAs. He was given by propranolol 5 mg once daily and referred for Blalock-Taussig (BT) shunt procedures.

Discussion: In this case, the pulmonary arterial supply was from multiple MAPCAs which further divide to supply all pulmonary segments on both sides. This might explain why our patient is unique as he did not develop cyanosis or breathlessness up to 3 years old due to MAPCAs supplying blood to lungs. The symptoms persisted might due to insufficient MAPCAs to preserve pulmonary blood flow (PBF). Thus, staged repair was planned for this patient, which comprises unifocalization of the MAPCAs followed by RV to PA continuity and closure of VSD.

Conclusion: BT shunt is necessary for patient with PA-VSD, especially in our patient who showed insufficient MAPCAs to preserve PBF. We should evaluate the MAPCAs before proceeding to definitive repair.

Biography

Evan Hindoro works as an Internship Doctor in Belitung Timur General Hospital. After graduating as a Medical Doctor in 2016, he embarked to enrich his skills and knowledge in Cardiology by working as a Research Assistant at the National Cardiovascular Centre Harapan Kita (NCCHK), Indonesia. Beside his responsibility as a Medical Doctor, he filled up his days with basic research training and participating in workshops. His credibility in medical research has been proven in several publications, both local and international journals. In 2015 his papers, titled "Fractional Flow Reserves: Nurturing a functional perspective in angioplasty" and "Routine thrombus aspiration in primary percutaneous coronary intervention: Is it still necessary?" have been published in European Heart Journal Supplement. He is currently working on a coronary artery diseases registry in the Departement of Cardiology, Siloam Hospital Lippo Karawaci, Tangerang, Indonesia.

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