

Patients misdiagnosed as persistent pulmonary hypertension of neonate are especially severe cases of total anomalous pulmonary venous connection

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Introduction: Total anomalous pulmonary venous connection (TAPVC) was occasionally misdiagnosed as persistent pulmonary hypertension of neonate (PPHN). Both disorder had severe pulmonary hypertension and interatrial right-left-shunt. If TAPVC patient is in particularly serious condition, its common chamber is hardly-detectable by echocardiogram. We predict PPHN-misdiagnosed patients were especially severe cases of TAPVC. We investigated clinical characteristics of PPHN-misdiagnosed patients with TAPVC. Methods: The medical records of six patients with TAPVC who was diagnosed as PPHN initially were reviewed. They underwent surgery between 1 day and 12 days after birth. We used 13 patients with TAPVC as control who underwent surgery between 1 day and 12d days. Clinical findings were compared between two groups. Results: In PPHN-misdiagnosed patients symptoms appeared within 4 hours after birth (83% vs. 7%, $p=0.0095$). Before surgery PPHN-misdiagnosed patients were more on respirator (100% vs. 7%, $p=0.00025$). In PPHN-misdiagnosed group inotropic drugs administered more (83% vs. 7%, $p=0.029$). Half of patients in PPHN-misdiagnosed group died after TAPVC repair, whereas only 15% died in correct-diagnosed group. The differences on images were two factors in PPHN-misdiagnosed group: pneumothorax (50% vs. 0%, $p=0.02$); small cardio-thoracic ratio 47% or less (66% vs. 7%, $p=0.021$). It is difficult to distinguish two groups in following factors: levels of oxygen saturation; degree of interstitial opacity; intensity of pulmonary hypertension; dimension of left ventricle. Types of TAPVC and existence of pulmonary venous obstruction are also not different between two groups. Conclusion: The patients misdiagnosed as PPHN were especially serious in cases where TAPVC repair was needed earlier after birth. They had smaller hearts on chest x-ray with pulmonary vein flow potentially decreased. Furthermore, they had pneumothorax. Possibly small common chamber were hard to be detected by echocardiography because of decreased pulmonary-vein flow and pneumothorax. There was no easy-to-understand information on images except for these two factors in PPHN-diagnosed patients. We should review infants to suspect TAPVC who were diagnosed as PPHN and medicated intensive care with small heart or pneumothorax. . .