

Systemic pulmonary hypertension in Down syndrome infants with ventricular septal defect is associated with hypoventilation and large various shunts

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Introduction: Systemic pulmonary hypertension (systemic PH) often resides in Down infants with ventricular septal defects (VSD). However, there are few reports to describe risk factors for systemic PH in Down infants with VSD. The purpose of this study is to investigate why systemic PH often subsists in Down infants with VSD. Methods: The medical records of 222 infants with VSD were reviewed (44 with Down syndrome and 178 with non-Down). All had no significant stenosis of pulmonary artery. They had cardiac catheterization in view of surgical intervention between 1993 and 2012. Arterial carbon dioxide pressure (PaCO₂) was measured during cardiac catheterization. Maximum dimension of VSD on echocardiography was calibrated by the body surface area. We calculated the right-to-left pressure ratio on end-systole point (RV/LV). We defined systemic PH as value of RV/LV in the top of fifth (RV/LV \geq 0.99). First, independent factors which influenced systemic PH were determined in all 222 infants. Second, we investigated relationship between these independent factors for systemic PH and Down syndrome group. Results: The ratio of patients with systemic PH was significantly higher in Down infants than in non-Down infants (38% vs. 16%). After multiple logistic regression analysis systemic PH in 222 infants was independently associated with large dimension of VSD (\geq 43mm/m²), existence of patent ductus arteriosus, existence of atrial septal defect/patant foramen ovale, and increased levels of PaCO₂ (\geq 47mmHg). The ratio of patients who had each factor was significantly higher in Down infants respectively. However, these four factors were not associated with systemic PH only in Down group, although they were all associated with systemic PH only in non-Down group. Discussion: This study revealed large VSD, accompaniment of other left-to-right shunts and hypoventilation were related to systemic PH, each of which more existed in Down infants. This would cause systemic PH more frequently in Down infants. However, these independent factors were not related to systemic PH only Down group, although those were associated with systemic PH only in non-Down group. Some other factor relevant to Down syndrome itself, which was not disclosed in this study, would be responsible for systemic PH.