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Cardio-pulmonary factors in patients with 22q11.2del are equal to those in patients with normal chromosome after repair of tetralogy of Fallot

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Background. The ratio of 22q11.2del syndrome is 15% in patients with tetralogy of Fallot (TOF). Patients with 22q11.2del often have abnormality of pulmonary arteries (PA), such as pulmonary atresia, major aorto-pulmonary collateral artery (MAPCA), and high pulmonary vascular resistance. Cardiac and pulmonary-artery complications arise in repaired TOF patients, particularly on remote period. However, there were no reports about cardio-pulmonary functions in repaired TOF patients with 22q11.2del. We investigate cardio-pulmonary matters in repaired TOF patients with 22q11.2del. Methods. The medical records of 121 repaired-TOF patients were reviewed aged from 1 to 53 years. We performed cardiac catheterization to grasp hemodynamic status between 2010 and 2015. Cardio-pulmonary performances were compared between patients with 22q11.2del and patients without chromosomal aberrancy. Results. We excluded 10 patients with abnormal chromosome other than 22q11.2del, the rest of whom we divided into two groups: 22q11.2 del group (n=11) and normal chromosome group (n=100). As type of PA in 111 patients pulmonary stenosis was 76, pulmonary atresia without MAPCA 24, and pulmonary atresia with MAPCA 11. The ratio of pulmonary atresia with or without MAPCA was different between 22q11.2del and normal chromosome (54% vs. 29%, $p=0.01$); that of Rastelli type surgery was also different between two groups (72% vs. 37%, $p=0.049$). Between 22q11.2del and normal chromosome, however, there were no significant differences in ratio of patients with severe PA valve regurgitation; reduced ejection fraction of RV; expanded RV volume; increased RV pressure. Similarly, estimations of pulmonary vascular bed were almost equal between two groups: mean pressure of PA, pulmonary vascular resistance, and pressure of pulmonary capillary wedge. As left ventricular factors, there were no significant differences between two groups. Clinically, the ratio of patients having symptoms was not different between two groups. Conclusion. Contrary to our exception, TOF patients with 22q11.2del possessed pulmonary vascularization equivalent to those with normal chromosome, if they had completed definitive repair. Similarly, they owned both ventricular functions which were not inferior to those in patients with normal chromosome. After radical surgery of TOF we might observe cardio-pulmonary problems of 22q11.2del in a similar way with normal chromosome.