

**Fontan patients with right isomerism possess inferior cardiac performances irrespective of strong valve regurgitation**

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**Background.**

When patients with right isomerism heart (Riso) possess intensive regurgitation of atrio-ventricle (AVVR), they suffer depressed cardiac functions. However, we often find Riso-patients to fall into cardiac impairment, although they have no strong AVVR. We investigated whether Riso-patients owned cardiac depression irrespective of intensive AVVR.

**Methods.**

The medical records of 36 Fontan patients with Riso were reviewed who underwent cardiac catheterization between 2010 and 2015. Control was 181 Fontan patients without Riso who had examinations likewise during the same period. First, we compared cardiac performances and clinical features between Fontan patients with and without Riso. Second, we excluded patients with AVVR mild-to-moderate or over (Riso 10, non-Riso 20) from 217 patients, using rest of whom we compared cardiac performances in the same way.

**Results.**

Riso-patients were operated for AVVR more (41% vs. 16%,  $p<0.001$ ); they had residual AVVR mild-to-moderate or over more (27% vs. 11%,  $p=0.0079$ ). As for cardiac performances, Riso-patients possessed larger ventricular volume on end-systole (64% vs. 51%,  $p=0.013$ ) and on end-diastole (125% vs. 106%,  $p=0.023$ ). They had higher capillary wedge pressure of pulmonary artery (9.1 vs. 6.9 mmHg,  $p=0.00082$ ). Driving pressure of pulmonary artery was lower in Riso-patients (3.7 vs. 5.3 mmHg,  $p=0.0016$ ). Between new Riso group ( $n=26$ ) and new non-Riso group ( $n=167$ ), where 30 patients with mild-to-moderate or over AVVR were excluded from origin groups, new Riso-patients also owned inferior cardiac performances: larger ventricular volume on end-systole (60% vs. 49%,  $p=0.042$ ) and end-diastole (120% vs. 103%,  $p=0.048$ ); more elevated capillary wedge pressure (8.5 vs. 6.7 mmHg,  $p=0.0034$ ); smaller driving pressure (3.7 vs. 5.6 mmHg,  $p=0.019$ ). In the background for new groups almost all Riso-patients had no left ventricle as major systemic pump (4% vs. 27%:  $p=0.00026$ ); they undertook shunting to pulmonary artery as 1st strategy which forcedly dilated ventricle. In addition, Riso-patients more undertook repair of atrio-ventricular valve regurgitation which had stretched ventricle before surgery.

**Conclusion.**

Our study showed Fontan patients with Riso possessed more dilated ventricle and higher capillary wedge pressure without reference to regurgitation of atrio-ventricular valve. Riso-patients potentially might have deteriorating cardiac function owing to inherent predisposition and pre-Fontan intervention.