

Reduced pulmonary diffusing capacity is highly increasable in Fontan patients

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Introduction

Fontan patients have reduced pulmonary diffusing capacity, however the cause remains unclear. We aimed to assess the etiology of this reduction, and in particular to distinguish between reduced alveolar capillary membrane diffusing capacity and/or reduced pulmonary capillary blood volume. Furthermore, we aimed to assess potential reversibility of the reduced diffusing capacity and to search for independent predictors of reduced diffusing capacity.

Methods

Advanced pulmonary function test were performed in the sitting position in 87 Fontan patients (mean age: 16.3 years, SD 7.6 years) using the single breath method. Of the 87 patients, 72 patients performed a symptom-limited cardiopulmonary exercise test. Before the exercise test cardiac index and stroke volume index were measured at rest using the inert gas-rebreathing method. Furthermore ten Fontan patients and nine healthy matched controls performed a supine pulmonary function test after a supine rest.

Results

The single breath test showed a mean diffusing capacity corrected for concentration of hemoglobin and alveolar volume (DLCOc/VA) of 70.3% of predicted, equal to a z score of minus 2.38. Alveolar capillary membrane diffusing capacity was normal (z score: minus 0.14) while pulmonary capillary blood volume was reduced (z score: minus 2.04). In a multiple linear regression analysis cardiac index at rest significantly predicted DLCOc/VA (regression coefficient: 0.18, $p < 0.001$). In the supine compared to sitting pulmonary function test, DLCOc/VA increased 51.8% in Fontan patients compared to 23.3% in the control group ($p < 0.001$) (Figure). Pulmonary capillary blood volume increased 48.3% in the Fontan group compared to 20.2% in the control group ($p = 0.001$) as well as alveolar capillary membrane diffusing capacity (14.1% in the Fontan group compared to 6.6% in the control group, $p = 0.008$).

Conclusions

We found a marked reduction in diffusing capacity in Fontan patients. In assessment of the etiology of this reduction we found a reduced pulmonary capillary blood volume while function of the alveolar capillary membrane appeared normal. The diffusing capacity was highly increasable in Fontan patients compared to a healthy control group, mainly due to increase in the pulmonary capillary blood volume. Cardiac index at rest was a highly significant independent predictor of reduced diffusing capacity.

