【Background】 Fontan patients are known to have augmented exercise ventilation. However, it is unsettled whether it is due to increased pulmonary dead space or augmented ventilatory drive. 【Methods】 Twenty-six Fontan patients, aged 8.5 ± 3.0 years, underwent symptom-limited treadmill cardiopulmonary exercise testing. Two groups of age- and sex- matched subjects served as controls: the biventricularly repaired (Bi, n=18), and the true control (C, n=29) groups. 【Results】 Peak oxygen uptake (VO2peak) was not different among groups (41.0 ± 8.4, 43.5 ± 6.6, and 45.9 ± 11.6 ml/min/kg for Fontan, Bi, and C groups, respectively, p=0.16). Fontan subjects, however, showed steeper alveolar ventilation/carbon-dioxide (VA/VCO2) regression slope (26.9 ± 3.4, 21.0 ± 2.0, and 21.6 ± 2.1, for Fontan, Bi, and C groups, respectively, p<0.001), as well as lower end-expiratory carbon-dioxide fraction (FetCO2VAT) at ventilatory threshold (VAT) (4.4 ± 0.5, 5.5 ± 0.5, and 5.5 ± 0.4%, for the Fontan, Bi, and C groups, respectively, p<0.001). The dead-space ventilation fraction at VAT was not different among the groups (0.33 ± 0.06, 0.33 ± 0.04, 0.35 ± 0.05 for the Fontan, Bi, and C groups, respectively, p = 0.54). Also in the Fontan subjects, arterial oxygen saturation at rest (SaO2rest) was correlated with VA/VCO2 regression slope (r =-0.41, p =0.04) and with FetCO2VAT (r =-0.53, p< 0.01). 【Conclusions】 Fontan patients are characterized by exercise hyperventilation due to augmented ventilator drive, which is further augmented by residual hypoxemia.