Exercise performance and quality of life is more impaired in Eisenmenger syndrome than in complex cyanotic congenital heart disease with pulmonary stenosis.

Abstract:
Patients with cyanotic congenital heart disease without corrective surgery or palliation survive into adulthood, if they have a balanced pulmonary blood flow facilitated by pulmonary stenosis (PS) or Eisenmenger syndrome (ES). Both groups show cyanosis, diminished exercise performance and impaired quality of life. This study aimed to compare the functional outcome of those two cohorts directly. In total fifty-eight cyanotic patients with cardiac shunts (28 male, 30 female, aged 14-55 years) were investigated, twenty-three of them with PS and thirty-five of them with ES. They completed the health related quality of life questionnaire SF-36 and performed a symptom limited cardiopulmonary exercise test. At exercise, oxygen saturation decreased severely and similarly in both groups (PS: 90% to 65% vs. ES: 87 % to 64%). Moreover, hemoglobin levels were comparable in both subgroups. Exercise capacity was markedly reduced, but more diminished in ES (PS: 20.3 (11.9; 24.6) ml/min/kg vs. ES: 11.3 (9.7; 14.5) ml/min/kg; p< 0.001) and ventilatory inefficiency expressed as V(E)/V(CO2) slope was more enhanced in ES (PS: 45.7 (37.6; 52.9) vs. ES: 54.6 (43.4; 68.7); p = 0.005). Oxygen saturation at rest was correlated to peak V(O2) (r = 0.436; p = 0.001) and V(E)/V(CO2) slope (r = -0.388; p = 0.003). Self estimated quality of life was poor, with worse results in physical and psychosocial domains in ES group. Despite similar
cyanosis, patients with ES show less exercise performance, more ventilation-perfusion-mismatch and a worse quality of life compared to complex cyanotic congenital heart disease patients with PS. Moreover, oxygen saturation at rest predicts exercise capacity and ventilatory efficiency in this cohort.