Impact of Restrictive Lung Disease on Morbidity and Mortality in Adults with Congenital Heart Disease

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Background: Compared to the general population, adults with congenital heart disease (CHD) have a higher rate of hospitalization and mortality. Reduced exercise capacity is also more common in adults with CHD, and identifies those patients with increased risk for hospitalization and death. Decreased exercise capacity in this population is often attributed to an underlying cardiac etiology, including chronotropic incompetence and cardiac dysfunction. However, recent studies have also reported a high prevalence of abnormal pulmonary function with restrictive lung physiology, that may also contribute to exercise intolerance. The impact of pulmonary function on clinical outcomes and mortality in adults with CHD is currently unknown. The aim of this study was to explore the relationship between pulmonary disease and the risk for hospitalization and death in adult patients with CHD.

Methods: Based on previous pilot data, we identified four patient populations with the highest prevalence for abnormal and restrictive lung function based on low forced vital capacity (FVC) percent predicted measured with spirometry: tetralogy of Fallot (TOF), congenital pulmonary valve stenosis (PVS), transposition of the great arteries status post atrial switch procedure (TGA), and functional single ventricle status post Fontan procedure (SV). Patients with any of the above diagnoses who underwent at least one measurement of lung function with spirometry at the time of cardiopulmonary exercise testing (CPET) between January 1, 2000 and March 31, 2014 were included in the study. Pulmonary function was classified based on FVC percent predicted as normal if > 70%, mildly reduced if 60-70%, and moderate-to-severely reduced if <60%. Assessment of the combined clinical outcome of death and/or non-elective hospitalization for primary cardiac or respiratory indication was obtained with retrospective chart review.

Results: A total of 292 total patients were included. Primary diagnosis was TOF in 143 (49%) patients, SV in 67 (23%), TGA in 46 (16%), and PVS in 36 (12%). The average age at spirometry testing was 27.1 ± 10 years. Pulmonary function was normal in 194 (66%) patients, mildly reduced in 57 (20%), and moderate-to-severely reduced in 41 (14%). Moderate-to-severely reduced pulmonary function was associated with greater number of previous cardiac surgeries ($p<0.001$), greater number of previous thoracotomies (<0.001), and diagnosis of scoliosis ($p=0.001$). Patients with moderate-to-severely reduced pulmonary function were also more likely to have abnormal CPET results including peak VO2 < 80 percent predicted ($p<0.001$), breathing reserve < 20% ($p<0.001$), and VE/VCO2 slope greater than 40 ($p=0.04$). During an average follow-up interval of 3.53 ± 2.9 years, 67 patients reached the combined clinical outcome of at least 1 non-elective hospitalization for primary cardiac or respiratory indication (n=59) and/or death (n=8). Utilizing a multivariable Poisson regression model, moderate-to-severely reduced pulmonary function was a significant and independent predictor of the combined clinical outcome of hospitalization and/or death (incidence ratio=3.16, 95%CI: 1.66-6.01, $p=0.002$).

Conclusion: Abnormal pulmonary function characterized by reduced FVC measured by spirometry is common in adults with CHD, and is associated with a greater number of previous cardiac surgeries and thoracotomies. Patients with moderately or severely reduced pulmonary function had a 3 times higher rate of hospitalization and/or death compared to those with normal or mildly reduced pulmonary function. FVC provides prognostic information in adults with CHD, and interventions to improve pulmonary function, such as pulmonary rehabilitation, should be studied in this population to potentially improve exercise capacity and reduce hospital admissions.