Lung Function in Patients With Congenital Heart Disease. Prevalence, Severity and Correlation to Diagnosis

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Objectives: Since restrictive lung function is an independent predictor of mortality in congenital heart disease (CHD), investigation of lung function in patients with CHD was performed; including description of prevalence, type of lung disease and severity and its correlation with the underlying diagnosis.

Methods: 536 patients with CHD aged 6 to 69 years (mean 28.2 ±13.4 years) underwent spirometry between 2012 and 2015. The ratio of forced expiratory volume in the first second to forced vital capacity (FEV1/VC Ratio) and vital capacity were measured. Cut-off: restrictive lung function: VC <80% of predicted, obstructive lung function FEV1/VC <0.8. Patients were divided in groups by diagnosis (see figure 1).

Results: Patients with normal lung function: 32.5%, restrictive lung function: 29.3%, obstructive lung function: 23.3%, mixed-type impairment: 14.9%. Highest percentage of restrictive lung defects was found in patients with complex cyanotic heart defects, univentricular anatomy, and Fallot tetralogy. Least patients with obstructive lung defect were found in patients with TOF and TGA after atrial switch operation (13.8 and 14.6%), whereas all other groups contain more than 22% of patients with obstructive lung disease. A decreased vital capacity correlates significantly with the number of thoracotomies (r=-0.41; P= 0.0001). There is a significant difference in lung impairment related to underlying diagnosis P<0.0001.

Conclusion: A significant amount of our patient population demonstrated impaired lung function. Restrictive lung function, detected in more than 40% patients with complex cyanotic anatomy, univentricular hearts and Fallot tetralogy may significantly affect the long term outcome. Since this is a known and independent risk factor for mortality (Alonso-Gonzalez, et al. 2013), it is recommendable to routinely perform spirometry especially on these patients to determine individual risk by impairment or decline in lung function.

Figure 1: Median and quartiles of VC% in different CHD. TGA – transposition of great arteries, ASO arterial switch operation, TOF – Fallot tetralogy.