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Impaired pulmonary function and its association with clinical outcomes, exercise capacity and quality of life in children with congenital heart disease

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Background: Impaired pulmonary function is an independent predictor of mortality in adult congenital heart disease (CHD), but has been scarcely studied in the paediatric CHD population. This study aims to compare the pulmonary function of children with CHD to healthy controls, and evaluate its association with clinical outcomes, exercise capacity, and quality of life.

Methods: Cross-sectional multicentre study among 834 children (555 CHD and 279 control subjects) who underwent a complete spirometry and a cardiopulmonary exercise test (CPET). The 5th centile (Z-score = -1.64) was used to define the lower limit of normal. The association of clinical and CPET variables with spirometry was studied using a multivariate analysis. Children and their parents filled in the Kidscreen health-related quality of life questionnaire.

Results: Forced vital capacity (FVC) and forced expiratory volume in one second (FEV1) Z-scores values were lower in children with CHD than controls (-0.4 ± 1.5 vs. 0.4 ± 1.3, P<0.001 and -0.5 ± 1.4 vs. 0.4 ± 1.2, P<0.001, respectively), without any obstructive airway disorder. Restrictive pattern was more frequent in CHD patients than in controls (20% vs. 4%, P<0.0001). FVC Z-scores were predominantly impaired in complex CHD, such as heterotaxy (-1.1 ± 0.6), single ventricle (-1.0 ± 0.2), and complex anomalies of the ventricular outflow tracts (-0.9 ± 0.1). In multivariate analysis, FVC was affected by the age, the body mass index, the maximum oxygen uptake, the genetic anomalies, the number of cardiac surgery and cardiac catheter procedures. FVC and FEV1 correlated with self and proxy-related quality of life scores.

Conclusion: These results suggest that pulmonary function should be monitored early in life, from childhood, in the CHD population.