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Original Article

Lung function test in children with left-to-right shunt congenital heart disease

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Abstract

Background Increased pulmonary blood flow may lead to abnormal lung function in children with left-to-right (L to R) shunt congenital heart disease. This condition has been linked to considerable mortality and morbidity, including reduced lung function.

Objective To assess for lung function abnormality in children with L to R shunt congenital heart disease.

Methods We conducted a cross-sectional study involving children aged 5-18 years and diagnosed with L to R shunt congenital heart disease at Dr. Sardjito Hospital from March to May 2017. Subjects underwent spirometry tests to measure forced expiratory volume-1 (FEV-1), forced vital capacity (FVC), and forced expiratory volume-1 (FEV-1)/forced vital capacity (FVC).

Results Of 61 eligible subjects, 30 (49.2%) children had atrial septal defect (ASD), 25 (41%) children had ventricular septal defect (VSD), and 6 (9.8%) children had patent ductus arteriosus (PDA). Spirometry revealed lung function abnormalities in 37 (60.7%) children. Restrictive lung function was documented in 21/37 children, obstructive lung function in 11/37 children, and mixed pattern of lung function abnormality in 5/37 children. Pulmonary hypertension was found in 21 children. There was no significant difference in lung function among children with and without pulmonary hypertension (P=0.072).

Conclusion Abnormal lung function is prevalent in 60.7% of children with L to R shunt congenital heart disease, of which restrictive lung function is the most common. There was no significant difference in lung function among children with and without pulmonary hypertension. **[Paediatr Indones. 2018;58:165-9; doi: http://dx.doi.org/10.14238/pi58.4.2018.165-9]**.

Keywords: *congenital heart disease; L to R shunt; spirometry; lung function*

Congenital heart disease (CHD) is a significant health problem, with the highest birth prevalences seen in low- and middle-income coountries.[1,2] Left-to-right shunt CHD results in increased pulmonary blood flow (Qp) beyond the regular systemic blood flow (Qs).[3] The pathophysiological changes depend on the size of the defect and volume of pulmonary blood flow (Qp), which have been associated with increasing pulmonary extracellular fluid. Other factors affecting the magnitude of shunts include defect location, patient age, and pressure gradient between the two chambers of the shunt.[4,5] Lung function abnormality in children with CHD are due to either structural impact on the airways, or abnormal pathophysiological mechanisms leading to increased lung fluid and/or significant pulmonary disease.[6]

Children with CHD are at greater risk of infection including those of the respiratory tract.[7] Respiratory problems are linked to considerable morbidity and

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mortality in children.[6] A previous study reported that moderate to severe impairment of lung function was an independent predictor of mortality among adults with CHD [hazard ratio (HR) 1.63; 95% confidence interval (CI) 1.01 to 2.63; P=0.004].[8] However, there has been limited study on lung function abnormality in children with (CHD).[9] We aimed to assess lung function abnormalilty in children with L to R shunt congenital heart disease.

Methods

This cross-sectional study involved children aged 5 to <18 years and diagnosed with L to R shunt congenital heart disease at Dr. Sardjito Hospital from March 1 to May 31 2017. Congenital heart disease was confirmed using echocardiography. We included all children who had never undergone definitive treatment of the defect. Subjects underwent spirometry testing. Children with acute and chronic disorders affecting spirometry performance, such as vomiting, vertigo, hemoptysis, chronic lung disease, recent eyes, or abdominal or thorax surgery were excluded. We obtained informed consent from all subjects' parents.

Demographic data, history of medical treatment, and echocardiography results were collected from medical records. Spirometry was performed using a vitalograph spirometer pneumotrac type 6800 to assess FEV1, FVC, and FEV-1/FVC. Spirograms were accepted if they were free from artifacts, had good starting effort and showed satisfactory exhalation.[10] Lung function abnormalities were classified following the plot (**Figure 1**).

Data were analyzed using *SPSS version 22.0* for Windows 2007, and presented as mean, median, or proportion, as appropriate. Chi-square test was used to analyze the results when comparing proportions. A probability value of less than 0.05 was considered to denote statistical significance. This study was approved by the Medical and Health Research Ethics Committee of Universitas Gadjah Medical School/Dr. Sardjito General Hospital, Yogyakarta, Indonesia.

Results

Of 61 eligible subjects, 30 (49.2%) children had ASD, 25 (41%) children had VSD, and 6 (9.8%) children had PDA (**Table 1**). Lung function abnormality was observed in 37 (60.7%) patients. Restrictive lung function was found in 21 (56.8% of those with lung function abnormality) patients, consisting of 14/21 mildly restrictive, 4/21 moderately restrictive, and 3/21 (14.3%) severely restrictive lung function abnormalities.

Eleven (29.7%) patients had obstructive lung function, out of which 90.9% had mild obstructive lung fuction and 9.1% had moderate obstructive lung function. Mixed type lung function abnormality was found in 5/37 patients.

Seventeen of 30 (56.7%) children with ASD had lung function abnormality. In the VSD group, lung function abnormality was found in 15/25 (60%) children. Of children with PDA, 5/6 (83%) had abnormal lung function, mostly of the restrictive type. **Figure 2** describes the prevalence of lung function abnormalities for each type of congenital heart disease.

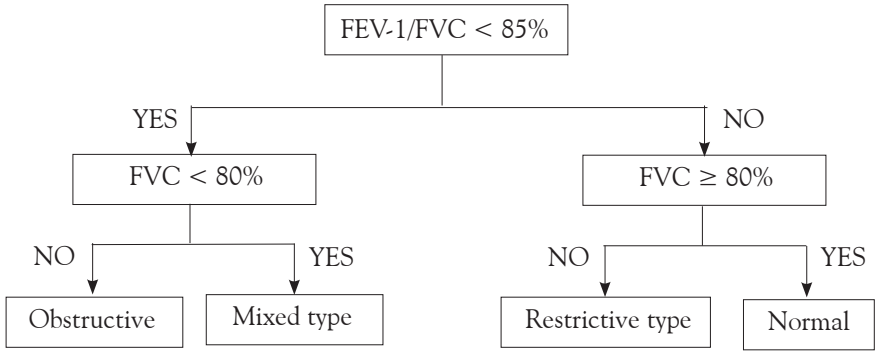

Figure 1. Interpretation of spirometry results[11]

Table 1. Baseline characteristics of subjects

Characteristics	(N=61)
Male sex, n (%)	28 (46)
Median age (range), years	10 (5-18)
Congenital heart disease, n (%)	
Atrial septal defect	30 (49)
Ventricle septal defect	25 (41)
Patent ductus arteriosus	6 (10)
Pulmonary hypertension, n (%)	21 (34)
Nutritional status, n (%)	
Good	37 (61)
Wasted	12 (20)
Severely wasted	11 (18)
Overweight	1 (1)

describes the results of the Chi-square analysis of lung function abnormality and pulmonary hypertension in our subjects.

Discussion

Defects at the great artery, atrial, or ventricular level which result in left-to-right shunts permit excess blood flow from the systemic circulation to the pulmonary circulation. This excessive pulmonary blood flow is linked to lung function abnormality in children with CHD.[5,7] Our study showed that 37/61 (60.7%)

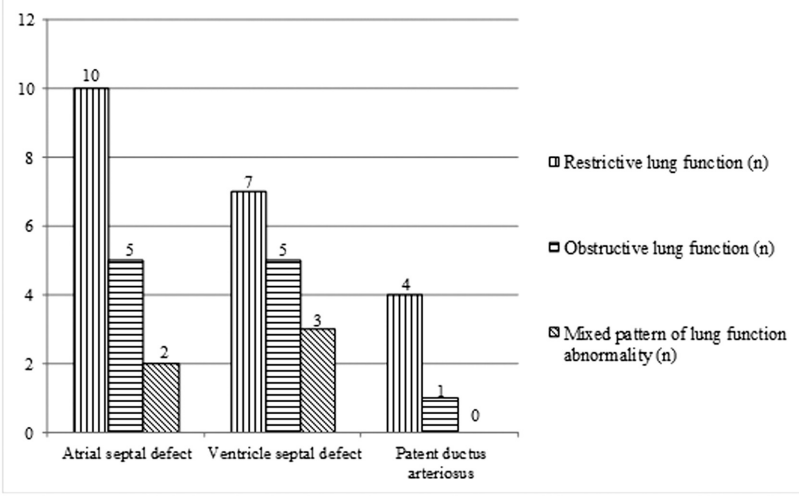

Figure 2. Lung function abnormality in each type of congenital heart disease

Based on echocardiography results, 21/61 left-to-right shunt CHD patients were diagnosed with pulmonary hypertension. The restrictive type was the most common lung function abnormality in subjects with pulmonary hypertension. There was no significant difference in lung function among children with and without pulmonary hypertension (P=0.072). **Table 2**

Table 2. Analysis of lung function abnormality and pulmonary hypertension in children with L to R shunt CHD

Pulmonary hypertension	Lung function, n (%)		P value
	Normal	Abnormal	
Yes	5 (23.8)	16 (76.2)	0.072
No	19 (47.5)	21 (52.5)	

children with CHD had lung function abnormalities. Yau *et al.* found that infants with CHD and left-to-right shunts had lower lung compliance and higher expiratory airway resistance than normal children (P<0.001).[12]

In our study, the most common lung function abnormality was the restrictive type, in 21 (56.8%) of subjects with lung function abnormalities. Similarly, Ginde *et al.* reported that restrictive lung function was prevalent in adults with CHD.13 Patients with a history of CHD tend to have mild to moderate restrictive changes, resulting in smaller lung volumes and flow rates relative to healthy subjects, leading to reduced ventilatory capacity, on average.[14]

Engorged vessels and volume-loaded heart chambers caused by left-to-right shunt CHD

may lead to external bronchial compression and obstructive lung function. Common sites of compression are the left main bronchus below the carina, which are compressed between an enlarged left atrium posteriorly and a dilated pulmonary artery or PDA anteriorly.[4,7] Partially obstructed airways develop a ball-valve effect and cause prolonged expiration. Completely obstructed airways lead to segmental atelectasis. The peak incidence is in infancy, when the bronchial cartilage is soft.[4] Only 11/61 children (18%) had obstructive lung function in this study. Lung function results may differ among age groups.

We found that 17/30 of patients with atrial septal defects had lung function abnormalities. This finding was consistent with a previous study in which 18/26 patients with ASD who had never undergone intracardiac correction had lung function abnormalities.[15]

Sari et al. performed spirometry in 20 children with VSD and found that 15/20 had restrictive lung function.[9] Similarly, 15/25 children with VSD had lung function abnormalities, of which 7/15 were the restrictive type. The larger the defect size, the higher the flow ratio. And more frequent respiratory tract infection in children with VSD increases the risk of restrictive lung function.[9]

Diagnosis of pulmonary hypertension in this study was established by echocardiography results. Echocardiography is used as a non-invasive method for assessing structural and functional intracardiac abnormalities, and monitoring disease progression over time. The sensitivity and specificity of echocardiography for pulmonary hypertension was 83% (95%CI 73 to 90) and 72% (95%CI 53 to 85), respectively.[16] A limitation of our study was that more accurate diagnoses of pulmonary hypertension should be done by right heart catheterization as the gold standard. Another limitation was not analyzing factors that may affect lung function abnormality in children with left-to-right shunt congenital heart disease.

In conclusion, abnormal lung function is prevalent in children with left-to-right shunt congenital heart disease, of which restrictive lung function is the most common. There is no significant difference in lung function among subjects with and without pulmonary hypertension.

Conflict of interest

None declared.

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