심폐재활

게시일시 및 장소 : 10 월 18 일(금) 08:30-12:20 Room G(3F)

질의응답 일시 및 장소: 10월 18일(금) 10:00-10:45 Room G(3F)

P 1-72

A relation between left ventricle ejection fraction and respiratory function in DMD patients

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Introduction

Duchenne muscular dystrophy (DMD) is a fatal disease caused by a mutation of dystrophin gene. It affects one in 4,000 male births and progressively results in muscle weakness, respiratory failure and heart failure. The lack of dystrophin impairs the connection between the cytoskeleton and the extracellular matrix, leading to progressive muscle weakness. In the heart, the degeneration of cardiomyocytes causes fibrosis and gradual decrease of systolic left ventricle ejection fraction (LVEF), eventually leading to dilated cardiomyopathy (DCMP) with heart failure. Proper and timely medical treatment of DCMP has been known to be crucial for survival in the DMD patients. Thus, accurate prediction and evaluation of cardiac function is important to proper treatment of DCMP in DMD patients. In DMD patients, respiratory function parameters are almost the only functional parameter that can be measured in advanced stage. Therefore, this study aims to determine if pulmonary function parameters can predict cardiac function in DMD patients.

Materials and Methods

Medical records of patients diagnosed with DMD admitted to the department of rehabilitation medicine were retrospectively collected, from January 1999 to March 2015. During admission, transthoracic echocardiogram (TTE) was performed to evaluate the cardiac function and various parameters of respiratory function, such as forced vital capacity (FVC), maximum insufflation capacity (MIC), maximal inspiratory pressure (MIP), maximal expiratory pressure (MEP), and peak cough flow (PCF) were also evaluated. We excluded the patients whose TTE and respiratory function test were not available. For patients with multiple admissions in the period, the only data for the first hospitalization period were included in the analysis. Among 179 patients, 72 were excluded due to missing TTE results. 7 patients were excluded because of insufficient TTE data. As a result,

data from 100 patients were qualified and finally analyzed. Pearson correlation and regression analysis were used to determine the degree of correlation.

Results

LVEF of the patients was 44.7 ± 16.2 %. And respiratory function parameters as follows; FVC 19.5 ± 14.6 %, MIC 1319 ± 511 mL, MIP 20.9 ± 17.9 %, MEP 13.3 ± 9.8 % and PCF 150 ± 58 L/min. On Pearson correlation study, various pulmonary function parameters (FVC, MIC, PCF, MIP and MEP) and LVEF showed no significant correlation on Pearson correlation (Table 1). And also, on simple linear regression analysis, respiratory function parameters (FVC, MIC, PCF, MIP and MEP) were not associated with LVEF (Table 2).

Conclusion

There was no statistically significant relationship between various pulmonary function parameters and LVEF. Therefore, heart failure in DMD, is only independent factor that has to be evaluated.

Table 1. Pearson correlation between LVEF and pulmonary functions in DMD patients

4	Correlation coefficient	P-value	
FVC	0.188	0.070	
MIC	0.028	0.785	
PCF	0.183	0.118	
MIP	0.096	0.350	
MEP	0.179	0.077	

LVEF; left ventricle ejection fraction, FVC; forced vital capacity, MIC; maximum insufflation capacity, PCF; peak cough flow, MIP; maximal inspiratory pressure, MEP; maximal expiratory pressure

Table 2. Association between LVEF and pulmonary functions (simple linear regression analysis)

8	LVEF		
	В	SE	P-value
FVC	0.212	0.116	0.070
MIC	0.001	0.003	0.785
PCF	0.054	0.034	0.118
MIP	0.087	0.093	0.350
MEP	0.295	0.165	0.077

LVEF; left ventricle ejection fraction, BMI; body mass index, B; β coefficient, SE; standard error, FVC; forced vital capacity, MIC; maximum insufflation capacity, PCF; peak cough flow, MIP; maximal inspiratory pressure, MEP; maximal expiratory pressure