

# 듀센형 근이영양증 환자에서의 심장 침범의 양상

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## Cardiac Involvement in Patients with Duchenne Muscular Dystrophy

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**Background** : Cardiac involvement in Duchenne muscular dystrophy (DMD) is common, but usually latent without symptoms or signs in the initial period of disease. This study investigated the incidence and predictor of cardiac involvement in DMD patients. **Method** : From January 2000 to June 2005, we enrolled 45 patients with DMD (aged 20.2 ±3.0 years) who admitted to the Yongdong Severance Hospital. Electrocardiography and transthoracic echocardiography was done to evaluate the cardiac function. **Result** : Electrocardiographic abnormalities were present in 80.1% of patients. Sinus tachycardia was most common (50%). LVEF was decreased (46.7 ±13.8%), and 56% of the patients had diastolic dysfunction. Patients with pulmonary involvement were older (20.7 ±3.8 vs 17.6 ±2.8 years, p=0.028), and patients with reduced LVEF (<50%) had longer duration of disease (11.4 ±4.4 vs 14.3 ±2.4 years, p=0.04). However, on multivariate analysis, age, duration of disease, pulmonary involvement, dyspnea symptom, electrocardiographic abnormality was not an independent predictor for LV systolic dysfunction in adolescent and adult patients with DMD. **Conclusion** : Cardiac involvement in adolescent and adult patients with DMD was frequently observed independent of age, duration of disease, pulmonary involvement, and dyspnea symptom. Therefore, more active cardiac investigation is required in patients with DMD, even without clinical suspicion.

**KEY WORDS** : Duchenne muscular dystrophy · Cardiac involvement · Echocardiography · Electrocardiography.

### 서론

2)3)  
80~85% 5  
Gower  
3500 1 가 , 10 가 가  
가 , 20  
1) 21 X 4)5)  
(short arm) (dystrophin)  
subsar- (20%). 6)7)  
colemmal cytoskeletal network 가 가  
actin filament dystroglycan complex 8)9)  
: 2005 10 14 . 10  
: 2005 12 14 , 18  
: , 135 - 270 146 - 92 . 10)  
: (02) 2019 - 3310, 2348 · : (02) 3463 - 3882  
E - mail : sejoong@yumc.yonsei.ac.kr 가 , 11)12) 가

가

13)

**대상 및 방법**

연구대상 및 연구방법

2000 1 2005 6

15

(209 ),

45

통계 분석

± , SPSS(Statistical Package for Social Science, SPSS Inc, Chicago, IL, USA) for windows, version 11.0 independent t-test, Chi - square test,

BNP, CK, p 0.05

CK - MB, TnT), 가 , 95%

심장기능 평가

, QRS

(two - dimensional),

(pulsed - wave, continuous - wave),

(color Doppler imaging)

호흡기능 평가

(82.6%)

(67.4%)

(capnography)

(nocturnal polysomnography)

45 mmHg

(hypoventilation)

(desaturation) 가 4%

가 50 mmHg

가 90% 가 5

통계 분석

± , SPSS(Statistical Package for Social Science, SPSS Inc, Chicago, IL, USA) for windows, version 11.0 independent t-test, Chi - square test,

p 0.05

가 , 95%

**결 과**

**대상 환자의 임상적 특징**

209

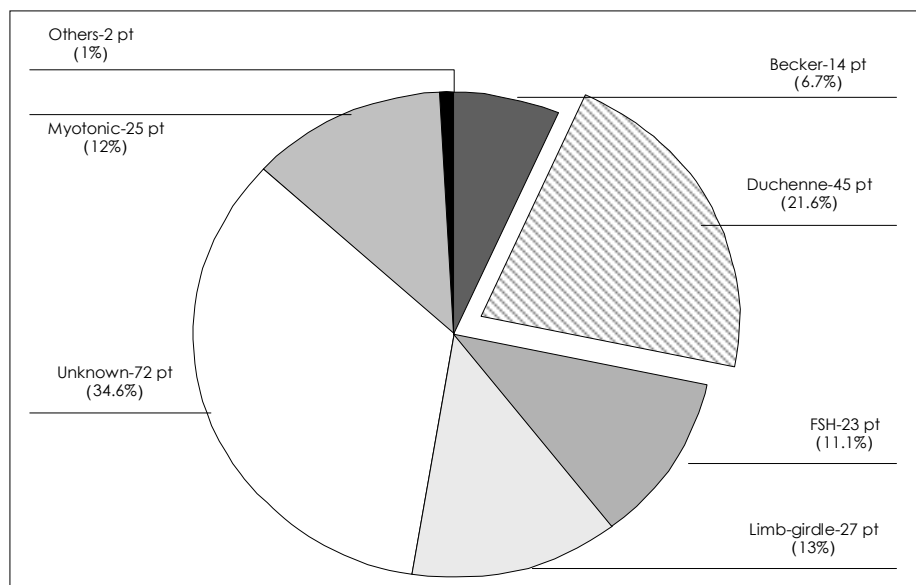
45 (21.6%) (Fig. 1).

45 20.2 ± 3.8 (15~33 ) ,

12.1 ± 4.2 (1~21 ) . 38

(82.6%) , 31

(67.4%) . , BNP 59.4 ± 137.3 pg/mL



**Fig. 1.** Frequencies of muscular dystrophy admitted to Yongdong Severance Hospital. FSH : Facioscapulohumeral muscular dystrophy.

CK 1101.4 ± 935.9 U/L 가 (Table 1).  
 42 (80.1%), 21 (50%), 7 (15.5%)  
 R (posterior R wave),  
 Q (inferior Q wave), Q (lateral Q wave)  
 가 3 (6.7%), 6 (13.3%), 16 (35.5%)  
 (Fig. 2, Table 2).

환자의 심초음파 소견  
 33 (51.6%)  
 46.7 ± 13.7%  
 2, 1  
 33  
 가 16 (56%)  
 (Fig. 3, Table 3).

**Table 1.** Baseline clinical characteristics of Duchenne muscular dystrophy patients

Sex (male %)	100
Age (yr)	20.2 ± 3.8
Duration of disease (yr)	12.1 ± 4.2
Ht. (cm)	155.7 ± 10.7
B.wt. (kg)	43.5 ± 13.4
BMI (kg/m <sup>2</sup> )	20.7 ± 5.2
Pulmonary involvement (%)	82.6
Presence of dyspnea (%)	67.4
PR (bpm)	93.1 ± 13.4
BNP (pg/mL)	59.4 ± 137.3
CK (U/L)	1101.4 ± 935.9
CK-MB (ng/mL)	22.6 ± 25.9
TnT (ng/mL)	0.023 ± 0.021

Data are mean ± SD, or number and percentage of patients. BMI : body mass index, BNP : B-type natriuretic peptide, B.wt. : body weight, CK : creatine kinase, Ht. : height, PR : pulse rate, TnT : troponin T

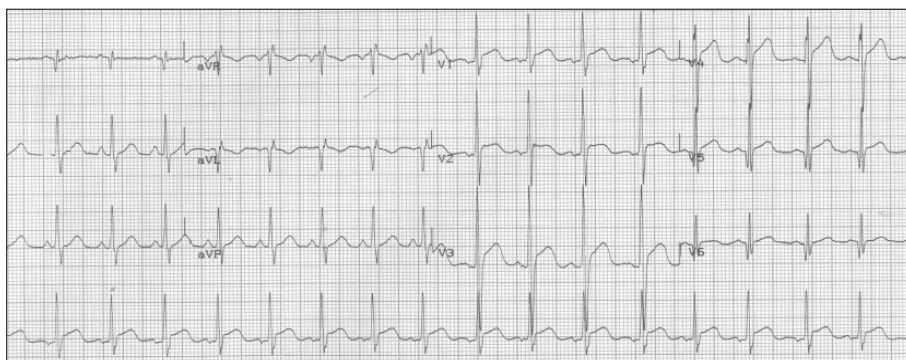
좌심실 수축기능 이상과 관련되 특성  
 (r = - 0.220, p=0.339)(Fig. 4),  
 (r = - 0.356, p=0.058)(Fig. 5).

50%  
 가  
 (11.4 ± 4.4 vs 14.3 ± 2.4 , p=0.04)(Table 4).  
 가  
 (19.4 ± 3.2 vs 21.2 ± 3.3 ),  
 BNP (9.1 ± 7.1 vs 89.6 ± 191.7 pg/mL).  
 (Table 4).

**Table 2.** Electrocardiographic abnormalities in Duchenne muscular dystrophy patients

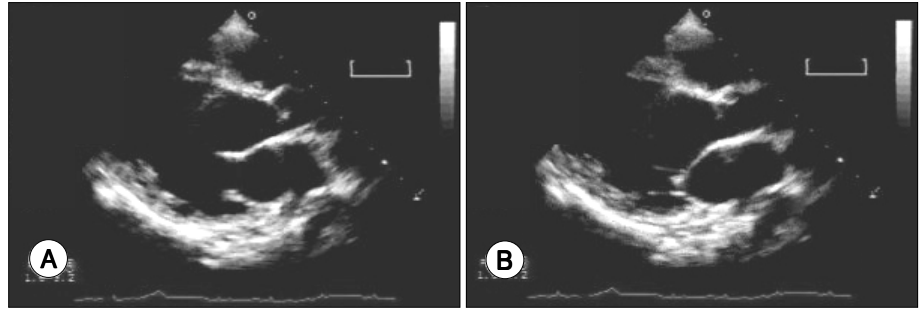
EKG abnormality	Patients No.(%)
Rhythm disturbance	
Sinus tachycardia	50.0
APCs	2.4
Ectopic A.tachycardia	0
AF/AFL	0
Conduction disturbance	
RBBB	16.7
LAFB	2.4
LBBB	0
AV block	0
VF/VT	0
Changes in QRS complex	
Tall R in V1 - 2	7.1
Inferior Q	14.2
Lateral Q	38.1

Data are number of patients. AF : atrial fibrillation, AFL : atrial flutter, APCs : atrial premature complex, A.tachycardia : atrial tachycardia, AV : atrioventricular, LAFB : left anterior fascicular block, LBBB : left bundle branch block, RBBB : right bundle branch block, VF : ventricular fibrillation, VT : ventricular tachycardia



**Fig. 2.** Twelve-lead electrocardiography in a patient with Duchenne muscular dystrophy shows sinus tachycardia (pulse rate 115 beats/min) with Q waves in lateral leads (lead I and aVL).

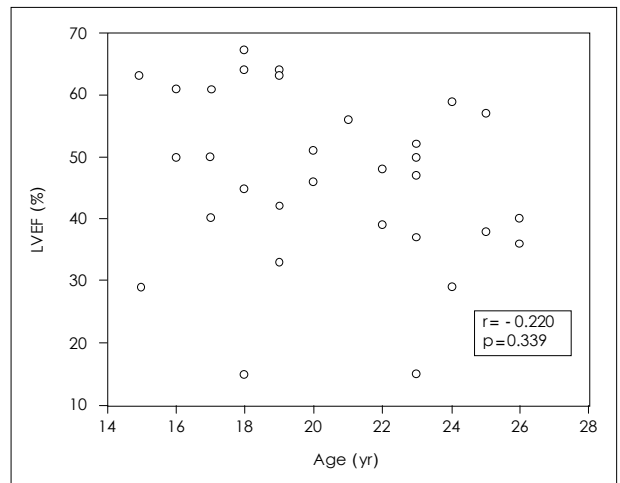
**Fig. 3.** Parasternal long-axis view on systole (A) and diastole (B) in a patient with Duchenne muscular dystrophy shows enlarged left ventricular chamber size (62 mm) with severe global hypokinesia of left ventricle (left ventricular ejection fraction=18%).



**Table 3.** Echocardiographic parameters in Duchenne muscular dystrophy patients

Parameter	
LVEF (%)	46.7 ± 13.8
LVEDD (mm)	46.8 ± 8.5
LA AP diameter (mm)	25.3 ± 7.6
Abnormal LV filling pattern	56.0%
Relaxation abnormality	2
Pseudonormalization	6
Restrictive LV filling pattern	1
E/E'	7.0 ± 1.8
Secondary valvular insufficiency	2
RWMA	1

Data are mean ± SD, or number and percentage of patients. AP : anteroposterior, E : E wave velocity, E' : E' wave velocity, LA : left atrium, LV : left ventricle, LVEDD : left ventricular end diastolic dimension, LVEF : left ventricular ejection fraction, RWMA : regional wall motion abnormality



**Fig. 4.** Relationship between age and left ventricular ejection fraction in Duchenne muscular dystrophy. LVEF : left ventricular ejection fraction.

폐 침범, 호흡곤란 유무에 따른 좌심실 수축기능 비교

가 (20.7 ± 3.8 vs 17.6 ± 2.8, p=0.028), BNP

가 (Table 5).

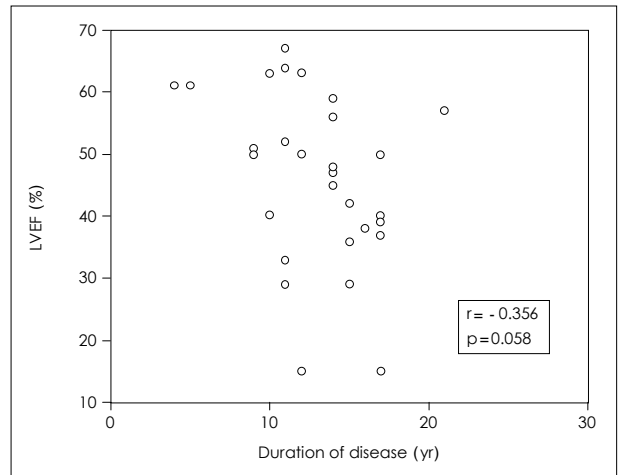
(47.1 ± 14.3 vs 43.0 ± 6.1%, p=0.28)(Fig. 6),

(49.0 ± 12.6 vs 46.1 ± 14.2%, p=0.637)(Fig. 7).

수축기 기능 장애의 예측 인자

가

(Table 6).



**Fig. 5.** Relationship between duration of disease and left ventricular ejection fraction in Duchenne muscular dystrophy (r = -0.356, p=0.058).

## 고찰

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가





