

## Product of Peak Systolic Wall Stress and Heart Rate Detects Preclinical Cardiomyopathy in Duchenne Muscular Dystrophy

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**Introduction:** Cardiomyopathy is a common late complication in Duchenne Muscular Dystrophy (DMD), but its onset is insidious and not clearly defined. We proposed "wall stress index (WSI)" as a marker of total left ventricular (LV) workload and tested whether it can detect preclinical myopathic changes in DMD.

**Methods:** Peak systolic wall stress (PSWS) was calculated in M-mode echocardiography with simultaneous measurement of systolic blood pressure (SBP). WSI was defined as PSWS x heart rate (HR). We measured WSI and LV mass index (LVMI) in normal controls, DMD with normal LV fractional shortening (FS  $\geq$  30%) (DMD-A), and DMD with decreased LVFS (< 30%) (DMD-B). The data were shown as mean  $\pm$  standard deviation.

**Results:** Total 36 normal controls and 47 DMD patients (83 DMD studies; 36 patients had two studies) were investigated. DMD-A was divided into two groups at the age of 10 years. HR was significantly higher in DMD groups than in controls. Despite comparable FS, WSI was significantly higher in DMD-A, even < 10 years, than in controls. WSI became even higher as FS further decreased (DMD-B). All DMD-B patients were treated with an angiotensin II-converting enzyme inhibitor and/or a  $\beta$ -blocker. In a longitudinal study with 33 DMD patients over 1 to 8 ( $4.0 \pm 2.0$ ) years, WSI showed a significant increase ( $p = 0.01$ ) with a decline in FS. LVMI remained mostly within normal limits but significantly increased with age ( $p = 0.021$ ).

**Conclusions:** Our data demonstrated that 1) WSI was significantly higher in young DMD than in controls despite comparable FS and that 2) WSI further increased as FS declined with age. The increase of WSI preceded the decrease in FS, suggesting its diagnostic value in detecting preclinical cardiomyopathy in DMD.

	Controls	DMD-A (< 10 yrs)	DMD-A ( $\geq$ 10 yrs)	DMD-B
Number	36	23	28	32
Age (yrs)	13.2 $\pm$ 2.9	7.1 $\pm$ 1.7	14.1 $\pm$ 3.2	16.4 $\pm$ 3.7
HR	70 $\pm$ 16	92 $\pm$ 13*	91 $\pm$ 13*	88 $\pm$ 13*
SBP (mmHg)	103 $\pm$ 13	92 $\pm$ 13*	105 $\pm$ 12	101 $\pm$ 11
FS (%)	36.8 $\pm$ 3.5	35.9 $\pm$ 3.0	34.8 $\pm$ 4.3	23.0 $\pm$ 4.4*
WSI	295 $\pm$ 68	444 $\pm$ 123*	483 $\pm$ 132*	686 $\pm$ 213*††
LVMI	29.8 $\pm$ 8.8	35.0 $\pm$ 5.9	32.6 $\pm$ 5.9	34.2 $\pm$ 12.2

$p < 0.05$  compared with control\*, DMD-A (< 10 yrs)†, and DMD-A ( $\geq$  10 yrs)‡

### Longitudinal Follow Up of 33 DMD Patients (4.1 $\pm$ 2.0 years)

	Before	After	p value
Age (yrs)	11.7 $\pm$ 3.6	15.8 $\pm$ 4.2	
%FS	32.3 $\pm$ 6.0	28 $\pm$ 7.3	< 10 <sup>-4</sup>
WSI	507 $\pm$ 163	600 $\pm$ 206	0.01
LVMI	31.6 $\pm$ 9.5	35.0 $\pm$ 10.5	0.021