

Meeting abstract

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## 2080 Relationship of structural and functional cardiac abnormalities to respiratory status in Duchenne Muscular Dystrophy

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### Introduction

Duchenne Muscular Dystrophy (DMD), a progressive functional myopathy, is characterized by death from respiratory and/or cardiac failure in early adulthood. Optimal care of these patients requires surveillance of both cardiac and pulmonary function.

### Purpose

We hypothesized that due to cardiopulmonary interaction, abnormal pulmonary function in DMD is associated with structural and functional cardiac changes measured by cardiac MRI (CMR).

### Methods

Retrospective cross-sectional study of 40 DMD subjects evaluated from 2004–2006. Each patient underwent standard pulmonary function testing and CMR for clinical indications (mean = 9 months between studies). Collected data included FVC% predicted (FVC%, a marker of ventilatory capacity) and FEV1%; Ejection fraction (LVEF, RVEF), end-diastolic volume (LVEDV, RVEDV), and end-diastolic mass (LVEDM, RVEDM). Spearman rank correlations were performed between pulmonary and cardiac parameters.

### Results

The 40 subjects ranged in age from 8 to 26 years and represented all stages of disability (ambulatory to profound weakness). Abnormal ventilatory capacity (FVC% <80)

was observed in 28 patients (70%). There was a significant negative correlation between FVC% and both LVEDM ( $r = -.48$ ,  $p = .002$ ) and RVEDM ( $r = -.42$ ,  $p = .01$ ) for all subjects. For the subset of subjects with an abnormal ventilatory capacity, the decline in FVC% correlated with LVEF ( $r = 0.42$  ( $p = 0.02$ )) but did not reach significance in its relation to RVEF ( $r = .27$ ,  $p = .09$ ).

### Conclusion

In DMD, RV and LV mass data are associated with ventilatory capacity, while LVEF is associated with ventilatory capacity only after both are abnormal. Prospective assessment is indicated to determine if RV and LV mass data can reliably predict ventilatory capacity, and in so doing supplement pulmonary surveillance to improve quality and quantity of life in this population.