

POSTER PRESENTATION

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Effect of pulmonary function on right heart function in Duchenne muscular dystrophy patients

Sharath Subramanian^{1*}, Kan N Hor², Wojciech Mazur³, Suzanne Smart¹, Tam Tran¹, Nancy Halnon⁴, Michael D Taylor⁵, Linda Cripe², Subha V Raman¹

From 18th Annual SCMR Scientific Sessions
Nice, France. 4-7 February 2015

Background

Duchenne muscular dystrophy (DMD) is an x-linked neuromuscular disorder in which cardiopulmonary disease leads to death in the third to fourth decade of life. Diaphragm and myocardium suffer progressive damage. While abnormalities in pulmonary and left ventricular function have been independently described, the relationship between pulmonary function and the right ventricle has not been evaluated.

Methods

Thirteen males with DMD enrolled in an ongoing clinical trial underwent CMR with multislice short axis cine imaging, using either breathhold segmented steady-state free precession or real-time acquisition techniques. Offline analysis of right ventricular volumes and ejection fractions was performed by a single reviewer blinded to PFT results using endocardial contour delineation and Simpson's rule. Forced vital capacity (FVC%) was recorded from PFTs acquired within 10 weeks of CMR examination, and serum brain natriuretic peptide levels were measured. Subjects were classified into two groups based on having at least 80% predicted FVC.

Results

Of the 13 subjects aged 17.5 ± 6.5 years (mean \pm SD) who underwent CMR and concurrent FVC assessment, 11 were non-ambulatory and all subjects were on chronic oral corticosteroids at the time of imaging. Left ventricular ejection fraction (LVEF) was normal in the study group ($56.9 \pm 5.3\%$). The right ventricular indexed

stroke volume was reduced in subjects who had less than 80% predicted FVC compared to patients who had more than 80% predicted FVC (25.2 ± 4.9 vs. 32.4 ± 5.3 mL/m²; $p=0.026$, Figure). There was no significant difference in the LVEF between two groups (55.5 ± 5.2 vs. 58.1 ± 5.6 , mean \pm SD; $p=0.4$). There was no correlation between indices of right ventricular size and systolic function and BNP levels.

Conclusions

Variable RV systolic function and pulmonary function were demonstrated in boys with DMD and normal LV systolic function as assessed by CMR. Abnormal pulmonary function is correlated with reduced RV stroke volume in boys with DMD and preserved LV systolic

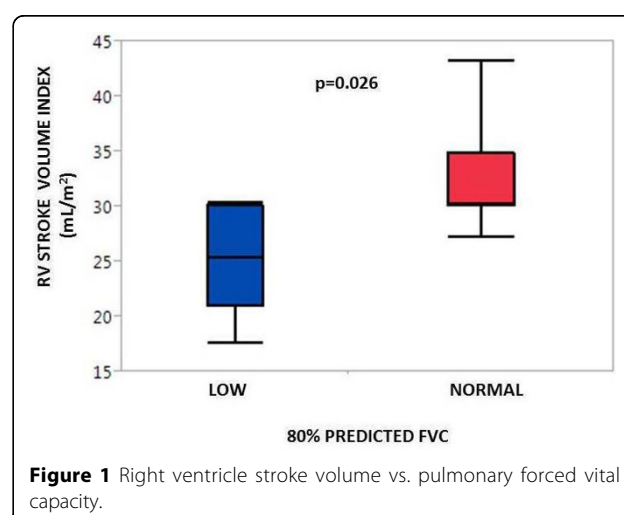


Figure 1 Right ventricle stroke volume vs. pulmonary forced vital capacity.

¹Cardiology, Ohio State University, Columbus, OH, USA
Full list of author information is available at the end of the article

function. Further studies are needed to ascertain the prognostic significance and the influence of ventilatory support on these findings.

Funding

This work was supported by BallouSkies.

Authors' details

¹Cardiology, Ohio State University, Columbus, OH, USA. ²Nationwide Children's Hospital, Columbus, OH, USA. ³The Christ Hospital Heart and Vascular Center, Cincinnati, OH, USA. ⁴University of California Los Angeles, Los Angeles, CA, USA. ⁵Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA.

Published: 3 February 2015

doi:10.1186/1532-429X-17-S1-P279

Cite this article as: Subramanian *et al.*: Effect of pulmonary function on right heart function in Duchenne muscular dystrophy patients. *Journal of Cardiovascular Magnetic Resonance* 2015 **17**(Suppl 1):P279.

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