Journal of Cardiovascular Magnetic Resonance

Poster presentation

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Early changes in right ventricular function and their clinical consequences in childhood dilated cardiomyopathy

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from 13th Annual SCMR Scientific Sessions
Phoenix, AZ, USA. 21-24 January 2010

Published: 21 January 2010

Journal of Cardiovascular Magnetic Resonance 2010, **12**(Suppl 1):P189 doi:10.1186/1532-429X-12-S1-P189

This abstract is available from: http://jcmr-online.com/content/12/S1/P189

Introduction

There is increasing evidence that the right ventricle (RV) plays a vital role in the clinical status in patients with dilated cardiomyopathy (DC) and their survival.

Purpose

1.) To determine whether the abnormalities in RV function that have been demonstrated in adults with dilated cardiomyopathy by X-ray ventriculography and echocardiography are also seen by magnetic resonance imaging (MRI) and in children with this disease. 2.) To investigate whether any alterations in RV size and function are clinically important.

Methods

We assessed biventricular size and function in eleven patients with dilated cardiomyopathy as well as in twelve normal paediatric controls. Late gadolinium enhancement images and phase contrast flow velocity mapping across both atrioventricular valves were obtained. Blood for N-terminal pro-brain natriuretic peptide (NT-pro-BNP) was collected at the time of MRI, and the results from the most recent echocardiogram and exercise test were reviewed.

Results

Patients with DC had significantly faster heart rates (85 versus 65 per minute), lower LV ejection fraction (EF) (42 versus 61%), RV EF (44 versus 54%) and lower LV and RV stroke volumes (35.5 versus 49.5 ml/m2 and 40.9 versus 56.4 ml/m2 respectively). Relative to the results in normal controls, RV EF was more severely reduced than LV EF (30.3 versus 18.7%). Patients had lower mitral and tricuspid valve inflow e/a wave velocity ratios than controls (2.02 versus 2.80 and 1.25 versus 2.58, respectively). The patient e/a wave ratios by MRI were not different from those derived by echocardiography. Tricuspid valve annulus velocity, measured by tissue doppler echocardiography, correlated with RV EF by MRI ($r = 0.60$, $p = 0.05$). RV EF and indexed right ventricular end-diastolic volume correlated with NT-pro-BNP ($r = -0.67$, $p = 0.03$, and $r = 0.65$, $p = 0.04$, respectively). RV EF correlated with the anaerobic threshold ($r = 0.67$, $p = 0.049$). Neither LV EF nor LV end-diastolic volume correlated with NT-pro-BNP or exercise tolerance. LV diastolic dysfunction, as shown by a lower MV e/a ratio correlated with lower LV EF($r = 0.90$, $p = 0.0062$), both measured by MRI.

Conclusion

Right ventricular systolic function is decreased in the early stages of DC and may, in fact, be more severely affected than the LV. The resulting decrease in stroke volume is compensated by an increase in heart rate to sustain a normal cardiac output. Right ventricular size and EF may be important indicators of subclinical heart failure and should be monitored by MRI. Phase contrast mapping of the atrioventricular valves can be used to assess diastolic function.