

Meeting abstract

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## 2084 Distinguishing between Fabry's disease and hypertrophic cardiomyopathy with early enhancement

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from 11<sup>th</sup> Annual SCMR Scientific Sessions  
Los Angeles, CA, USA. 1–3 February 2008

Published: 22 October 2008

*Journal of Cardiovascular Magnetic Resonance* 2008, **10**(Suppl 1):A353 doi:10.1186/1532-429X-10-S1-A353

This abstract is available from: <http://jcmr-online.com/content/10/S1/A353>

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### Background

CMR based pattern of late enhancement (LE) has been shown to differentiate between Fabry's disease, and hypertrophic cardiomyopathy. In Fabry's disease, LE is found at the lateral wall, while it is located at the septal insertion points of the right ventricle in hypertrophic cardiomyopathy. However, given that both conditions result in increased wall thickness, and in some patients, there is generalized LE, differentiation between the two cardiomyopathies becomes difficult. The value of CMR-based inflammatory markers such as edema (T2), and hyperemia (EE), as tools to further distinguish between these cardiomyopathies is not understood.

### Methods

We assessed 12 patients with Fabry's disease (5 males,  $42 \pm 18$  years) and 8 patients with hypertrophic cardiomyopathy (4 males,  $62 \pm 18$  years) using a 1.5 T MRI system (Avanto®, Siemens Medical Solutions, Erlangen, Germany). Left ventricular (LV) function and volumes were assessed using standard methods. Myocardial edema was imaged with a T2-weighted STIR sequence. Early contrast enhancement was quantified in free-breathing T1-weighted spin echo images before and early (over 4 minutes) after administration of 0.1 mmol/kg Gd-DTPA. Irreversible injury was visually assessed in images obtained by an inversion-recovery prepared gradient echo sequence 10 minutes after a total of 0.2 mmol/kg Gd-DTPA.

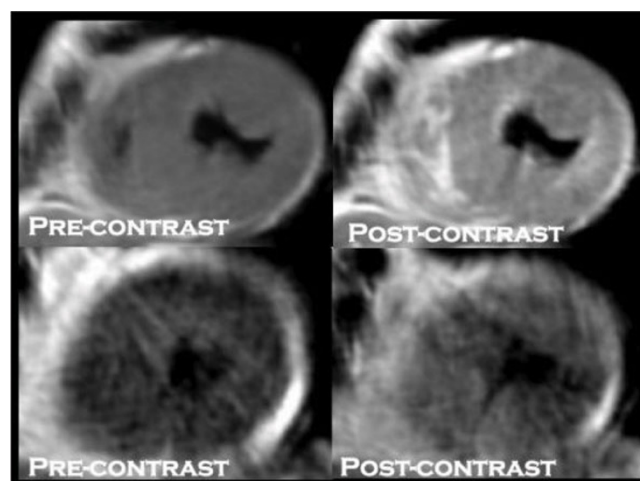
### Results

All patients had evidence of myocardial fibrosis. Measures of EE differed significantly between the two groups

(Fabry's disease  $6.85 \pm 2.96$ ; HCM  $3.64 \pm 0.97$ ;  $p < 0.05$ ). In contrast, there was no significant difference between T2 ratios ( $1.56 \pm 0.24$  vs.  $1.72 \pm 0.36$ ,  $p = \text{ns}$ ), LVEDV ( $133 \pm 37$  ml vs.  $141 \pm 43$  ml,  $p = \text{ns}$ ), LVESV ( $51 \pm 18$  ml vs.  $49 \pm 31$  ml,  $p = \text{ns}$ ), LVSV ( $82 \pm 21$  ml vs.  $91 \pm 17$  ml,  $p = \text{ns}$ ), nor LVEF ( $62 \pm 4\%$  vs.  $67 \pm 10\%$ ,  $p = \text{ns}$ ). Figure 1.

### Conclusion

Early contrast enhancement is increased in Fabry's disease and may help to differentiate this disease from other



**Figure 1**

Elevated early enhancement ration in a patient with Fabry's disease (upper panel) 4.68, compared to a ration of 2.24 in a patient with hypertrophic cardiomyopathy (lower panel).

causes of LV hypertrophy such as hypertrophic cardiomyopathy. Therefore, in addition to LE, EE should be performed in patients with hypertrophy of uncertain etiology.

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