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<td><strong>Author(s)</strong></td>
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Relationship between MRI derived right ventricular mass and left ventricular involvement in patients with anderson-fabry disease

Qin Li3,1, Ming-Yen Ng1,2*, Anna Calleja3, Djeven P Deva1,4, Andrew Crean1,3, Christiane Gruner3, Robert M Iwanochko3, Paaladinesh Thavendiranathan3,1

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

Background
Anderson-Fabry’s Disease is an X-linked lysosomal storage disorder in which there is an accumulation of globotriaosylceramide in both atria and ventricles as well as valvular tissue. We sought to investigate if there was a correlation on cardiac MRI between the degree of left ventricular (LV) involvement (ie. LV mass or late gadolinium enhancement) and right ventricular mass.

Methods
The study was approved by the research ethics board. Patients with Anderson-Fabry’s disease were identified through the Metabolic Genetic Disease Clinic between 1st January 2000 and 31st December 2013. We included only patients with genotyping and leucocyte alpha galactosidase A activity test confirming a diagnosis of Anderson-Fabry’s disease. Clinical data obtained included age and body mass index. Cardiac MRI was analysed using CMR 42 (Circle Cardiovascular Imaging) to measure LV and right ventricular (RV) volumes, ejection fraction and mass using short axis steady state free precession (SSFP) cine images (see figure 1). Presence of late gadolinium enhancement (LGE) and degree of LGE was calculated using a threshold of 6 SD above a remote region. Right ventricular hypertrophy (RVH) was defined as >28g/m² for males and >27g/m² for women. This threshold was based on a paper by Hudsmith et al (2005 Journal of Cardiovascular Magnetic Resonance).

Figure 1 Assessment of RV volume, function, and mass using short axis SSFP cine images (CMR42). The RV septal band was included in the assessment of the RV mass.

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Table 1 Comparison of LV mass and LGE quantification in Anderson-Fabry’s patients with increased and normal RV mass with p-values from unpaired t-test.

<table>
<thead>
<tr>
<th>LV mass (g/m²)</th>
<th>Increased RV mass</th>
<th>Normal RV mass</th>
<th>p-value</th>
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<tr>
<td>LGE 6SD (g)</td>
<td>12.48 ±24</td>
<td>1.44 ±2.29</td>
<td>0.0047</td>
</tr>
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Comparisons between RV mass, LV mass and LGE were performed using Pearson’s correlation and unpaired student t-test.

Results
43 patients (22 males) with a mean age of 45 ± 14yrs were included. There was good correlation between LV and RVEF (r=0.59, p<0.01), and LV and RV mass index (r=0.67, p<0.01). Total LV scar was higher in patients with right ventricular hypertrophy (RVH) (12.48g ± 24g) than in those without RVH (1.44g ± 2.29g) (p=0.0047). There was also a statistically significant difference between the LV mass of patients with increased RV mass (117.03g/m² +/- 69g/m²) compared to patients with normal RV mass (71g/m² +/- 28.7g/m²) (see table 1).

Conclusions
Patients with Anderson-Fabry’s disease with increased LV mass and LGE had a positive correlation with increased RV mass. RV mass changes appear to occur in more advanced myocardial involvement with Anderson-Fabry’s disease.

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Erratum: Relationship between MRI derived right ventricular mass and left ventricular involvement in patients with anderson-fabry disease

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Following publication of this abstract [1] it was noted by the authors that an incorrect version of Figure 1 had been inadvertently uploaded with the manuscript. The correct version Figure 1 is available below.

In addition it was found that the author list was incorrectly ordered, this has been corrected in the author list above.

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Figure 1 Assessment of RV volume, function, and mass using short axis SSFP cine images (CMR42). The RV septal band was included in the assessment of the RV mass.