

## BSCMR Abstract Submission 2019

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1 INFLAMMATORY CARDIOMYOPATHY IN FABRY DISEASE

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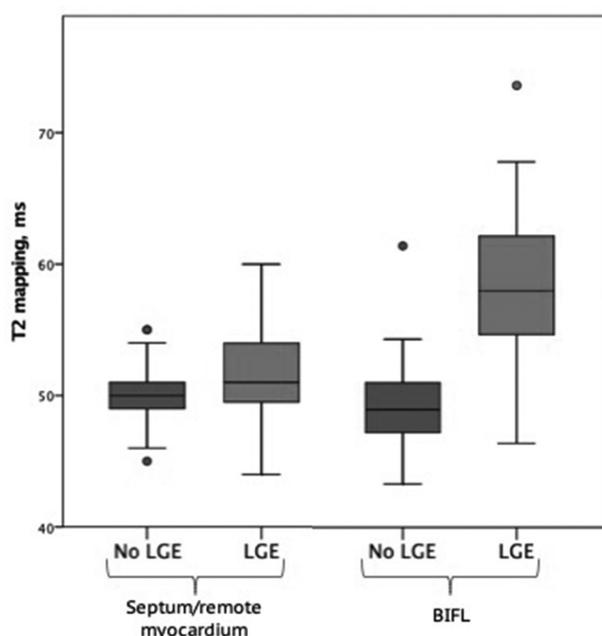
10.1136/heartjnl-2019-BSCMR.1

**Background** Fabry disease (FD) is an X-linked lysosomal storage disorder caused by mutations in  $\alpha$ -galactosidase A. Cardiovascular magnetic resonance (CMR) has helped unveil the pathogenesis of Fabry cardiomyopathy: sphingolipid storage (low T1 mapping values), left ventricular hypertrophy (LVH) and myocardial fibrosis with late gadolinium enhancement (LGE) characteristically present in the basal inferolateral (BIFL) wall. Recent evidence has suggested that the LGE may be inflammation and oedema as part of this pathogenic process.

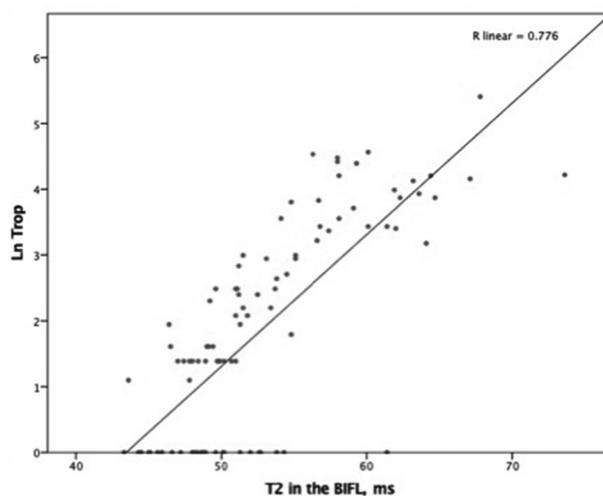
**Purpose** To assess the presence of inflammation in patients with FD using T2 mapping (for oedema/inflammation) supported by blood troponin levels (showing myocyte death and by inference inflammation).

**Methods** A multi-centre international study in gene positive FD patients using CMR and blood biomarkers. All participants underwent CMR at 1.5 T. Native T1 and T2 mapping were performed. The T1 mapping sequence was MOLLI with sampling scheme in seconds. LGE used a phase sensitive inversion recovery sequence. Global longitudinal 2D strain (GLS) values were obtained using feature tracking analysis. Blood high-sensitivity troponin T (hsTnT) was measured on the same day.

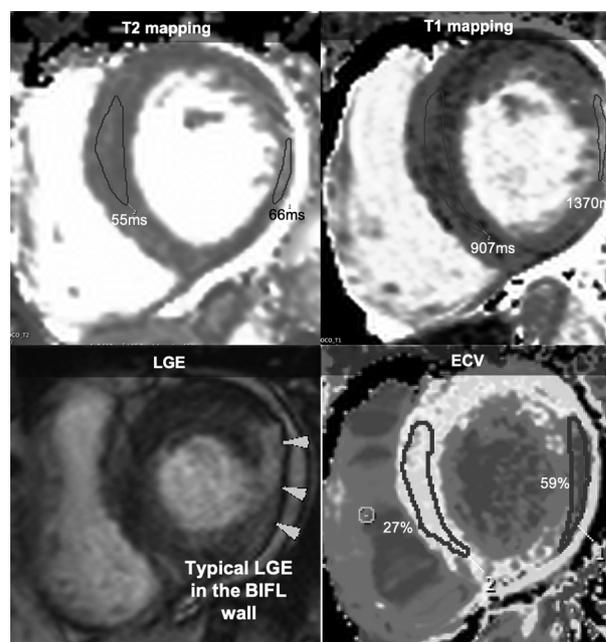
**Results** 100 FD patients (age  $43.8 \pm 1.3$  years, 42% male) were included. 45% had LVH, 35% LGE. Low T1 mapping



**Abstract 1 Figure 1** Box-plot graph showing T2 values in remote and BIFL (LGE) areas



**Abstract 1 Figure 2** Scatter-plot graph illustrating a positive correlation between T2 values in the BIFL wall and blood hsTnT



**Abstract 1 Figure 3** Native T1, T2 and ECV mapping values measured in the septum and basal inferolateral wall. Typical BIFL wall scar is seen. Note the increased T2 mapping values matching the areas of LGE

(normal  $< 943$  ms) was found in 49% and 33% had high hsTnT values (normal  $< 15$  ng/L). Mean T2 mapping values were  $52.6 \pm 0.6$  ms in the BIFL wall and  $49.5 \pm 0.3$  ms in the remote myocardium/septum ( $p < 0.001$ , normal  $< 53$  ms). T2 values in the BIFL wall were significantly higher among patients with LGE ( $58.2 \pm 6.1$  ms vs  $49.2 \pm 3.1$  ms,  $p < 0.001$ , figure 1). In a per-segment analysis of 1600 segments, higher T2 values correlated positively with percentage of LGE per segment ( $r = 0.262$ ,  $p < 0.001$ ), T1 values ( $r = 0.205$ ,  $p < 0.001$ ), maximum wall thickness ( $r = 0.253$ ,  $p < 0.001$ ) and GLS values ( $r = 0.212$ ,  $p < 0.001$ ). HsTnT values were higher among patients with LGE (median of 31 vs 3 ng/L in patients without LGE,  $p < 0.001$ ). There was a strong positive correlation between T2 values in the BIFL wall and  $\ln(\text{hsTnT})$  ( $r = 0.776$ ,  $p < 0.001$ , figure 2). The strongest predictor of increased hsTnT

in multivariate analysis (age, sex, LVH, septum T1, T2 in the BIFL, GLS, LGE) was T2 in the BIFL wall ( $\beta=0.4$ ,  $p=0.001$ ). **Conclusions** Cardiac involvement in FD goes beyond storage (low T1 values). When LGE is present, this is almost always associated with a high T2 and troponin elevation supporting FD as a chronic inflammatory cardiomyopathy. Initial reports of LGE being fibrosis are too simplistic – LGE in FD appears to have a significant chronic inflammation/oedema component.

**2** **NEO-AORTIC DILATATION AND REGURGITATION DURING PREGNANCY FOLLOWING THE ROSS PROCEDURE: AN EVALUATION OF CARDIAC MAGNETIC RESONANCE (CMR) DATA**

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**Background** The neo-aorta produced by the pulmonary autograft following the Ross procedure has excellent long-term outcomes, with an increasing number of women now undergoing pregnancy following Ross. Cardiac Magnetic Resonance (CMR) has been used to evaluate the risk of progression of neo-aortic dilatation and dissection that may arise as a result of the additional cardiovascular risks posed during pregnancy. Although there have been reported cases of dilatation and dissection, there is little research.

**Aims** To evaluate the extent of neo-aortic dilatation, and associated aortic regurgitation, during pregnancy following the Ross procedure.

**Methods** A retrospective cohort of women were identified who had undergone a Ross procedure after 1985, who in January 2019 were aged >16, in an adult congenital cardiology and joint cardiac-obstetric centre. CMR data was used to evaluate neo-aortic root dimensions and aortic regurgitation extent

pre-, during and post- pregnancy. A control group of case-matched patients who had undergone the Ross procedure, but not pregnancy, was used for comparison.

**Results** In all, 8 women carried a total of 15 pregnancies to term. In only one pregnancy did the mother experience serious cardiac decompensation, necessitating early delivery and associated with subaortic stenosis, mild aortic root dilatation and increasing aortic regurgitation. Four women (50%) experienced an element of aortic root dilatation during pregnancy, with an average increase between them of 0.525 cm (range +0.4 to +0.7). In no case did this lead to progressive dilatation or dissection. When compared to the control group, pregnancy was found to carry an increased risk of neo-aortic dilatation (Mean aortic root change for group: +0.26 cm (SD 0.30) pregnancy vs. +0.16 cm (SD 0.27) control). There was a high rate of initial mild aortic regurgitation in both groups. In three women their extent of regurgitation increased post-pregnancy, correlated with increased parity and aortic dilatation. In the control group only two women experienced increased regurgitation, only one of which was associated with neo-aortic dilatation.

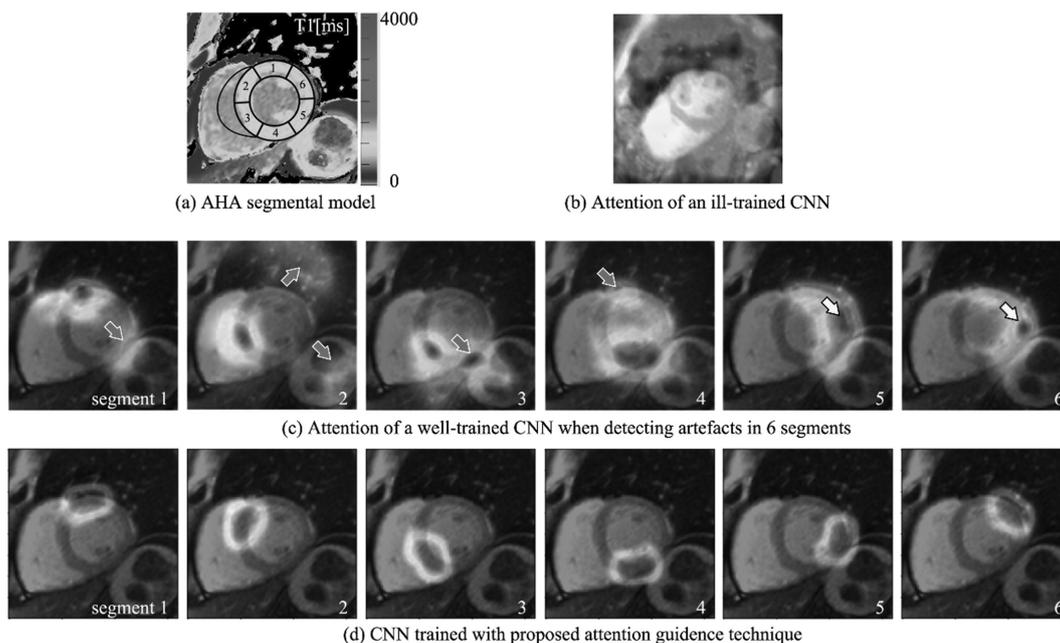
**Conclusions** Pregnancy appears generally well tolerated by women following the Ross Procedure, although there is an increased risk of neo-aortic dilatation and corresponding increase in aortic regurgitation compared to the control group.

**3** **TRAIN THE AI LIKE A HUMAN OBSERVER: DEEP LEARNING WITH VISUALISATION AND GUIDANCE ON ATTENTION IN CARDIAC T1 MAPPING**

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10.1136/heartjnl-2019-BSCMR.3

**Background** Artificial intelligence (AI) is increasingly used in diagnostic imaging. Deep convolutional neural networks (CNN)



**Abstract 3 Figure 1** Attention maps as the CNN ‘eye tracker’ in detecting T1-mapping artefacts in the 6 AHA segments (a), which reveals that (b) an ill-trained CNN looked at the features not desired for the task; In comparison; (c) a well-trained CNN correctly identified the segments, but with distraction (red arrows) and low accuracy (yellow arrows); (d) CNN trained with attention guidance looked at the target myocardial segments more accurately