Abstract 069 Table 1

Black ethnicity data	Afro-Caribbean patients N=170	All other patients N=972	p Value
Age	71 (53–77)	73 (63–81)	<0.01
Male gender	110 (64.7%)	652 (67.1%)	<0.01
Ischaemic cardiomyopathy	22 (12.9%)	406 (41.8%)	<0.01
Non-ischaemic (total)	148 (87.1%)	566 (58.2%)	<0.01
Amyloid (AL and TTR)	27 (15.9%)	21 (2.2%)	<0.01
ATTR V122I	17 (10%)	4 (0.4%)	<0.01
Dilated cardiomyopathy	46 (27.1%)	179 (18.4%)	<0.01
Hypertensive heart disease	32 (18.8%)	68 (7.0%)	<0.01

070 ENZYME REPLACEMENT THERAPY IMPROVES CARDIAC FEATURES OF FABRY DISEASE

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Introduction Fabry disease (FD) is an X linked deficiency of α -galactosidase A which leads to an accumulation of globotriaosylceramide (GL-3) throughout the body, particularly in the heart, brain and kidneys. Although left ventricular hypertrophy (LVH) in FD can improve with enzyme-replacement therapy (ERT), the response is difficult to predict. Furthermore, the response of other cardiac features such as aortic dilatation and ECG changes are poorly understood.

Methods A local registry of 66 patients with FD was studied. ECG, echocardiogram and Fabry Outcome Survey-Mainz Severity Score

Abstract 070 Figure 1 Effect of enzyme replacement therapy (ERT) on LV morphology and function in patients with LVH at baseline. Left ventricular mass index (LVMI), maximal wall thickness (MWT), LV end-diastolic diameter (LVEDD) and ejection fraction all showed significant improvements (all p values <0.05) at the end of follow-up (median 3 years). [Values shown are mean ± SD].

Index (FOS-MSSI) data were compared between baseline and after long-term ERT (median 36 months).

Results In patients with LVH (n=42), left ventricular mass index (LVMI), maximal wall thickness (MWT), left ventricular end-diastolic diameter (LVEDD) and ejection fraction (EF) were all seen to improve after ERT (LVMI: 135±13 vs 133±13 g/m², MWT: 17±6 vs 16±5 mm, LVEDD: 55±6 vs 54±6 mm; EF: 62±5 vs 64±3%; p<0.05) (Abstract 070 figure 1). In the entire patient group, PQ interval and P wave duration significantly increased with ERT (PQ: 131±13 vs 144±13 ms, P: 76±5 vs 90±6 ms; p values <0.001); QTc interval significantly decreased (418±18 vs 410±15 ms; p<0.001); and median FOS-MSSI score fell from 16 to 14 (p<0.001) (Abstract 070 figure 2). On logistic-regression analysis, none of the recorded baseline features (age, gender, LVMI, MWT, LVEDD, aortic diameter, EF, PQ interval, P wave duration, QRS duration, QT interval, Romhilt-Estes score or FOS-MSSI) predicted improvements in LVH or FOS-MSSI with ERT (p>0.05).

Abstract 070 Figure 2 Effect of enzyme replacement (ERT) on the conduction system. There was a significant increase in P wave duration and PQ interval from baseline; there was a significant decrease in QRS width and QTc interval from baseline at the end of follow-up (median 3 years). [Values shown are mean ± SD].

Conclusions ERT improved LV morphology and function in patients with LVH- but there was no relationship between age, gender, FOS-MSSI or baseline ECG/TTE features and the response. ERT also normalised long QTc intervals, short PQ intervals and short P waves; and reduced disease burden (FOS-MSSI).

071 CONCENTRIC HYPERTROPHIC REMODELLING AND SUBENDOCARDIAL DYSFUNCTION IN MITOCHONDRIAL DNA POINT MUTATION CARRIERS

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Background Cardiomyopathy is an independent predictor of morbidity and early mortality in mitochondrial DNA disease, and