instant feedback about their current health status; 2) Personalized advice: patients want a system that can adapt medication, sport activities and food recommendations to their current health status; 3) Transparency: patients want to know the source, reasons, and individualized interpretation of any recommended changes to the management of their condition. Additionally, patients have a desire to adapt their lifestyle to the needs of HF, but they require help to remain motivated to achieve this goal.

Conclusion These findings provide valuable information for the development and implementation of eHealth solutions. Patients want reassurance 24/7, independently of the availability of healthcare services, combined with personalized medical advice regarding day-to-day management of their HF. In a next step, we are planning a multicentre clinical trial to test the first prototype of the eHealth product (DoctorMe).

19 DYSTROPHIN DEFICIENT CARDIOMYOPATHY – PREDICTORS ASSOCIATED WITH THE CARDIAC PHENOTYPE IN A DUCHENNE’S REGISTRY POPULATION – A GUIDE FOR DEVICE THERAPY

¹J Carron, ¹SJ Fitzgerald, ¹M Egan, ²R Costello, ¹R Sheahan. ¹Department of Cardiology, Beaumont Hospital, Dublin, Ireland; ²Department of Respiratory Medicine Beaumont Hospital, Dublin, Ireland

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Background Patients with Duchenne muscular dystrophy (DMD) typically exhibit cardiac dysfunction and arrhythmia. With increasing life expectancy due to advances in respiratory support, cardiomyopathy and associated dysrhythmia are fast becoming the primary cause of morbidity and mortality in this patient group. Despite advances, the correlation between genotype and cardiac phenotype remains poorly understood and individual registries small, with implementation of device therapy often delayed due to poor diagnostic image quality.

Methods A single-center registry for DMD patients was established and data including genotyping, medical therapy and investigations such as cardiac MRI, nt-Pro BNP levels, echocardiogram and holter monitor was analysed. The aim was to potentially identify predictors associated with a more severe cardiac phenotype.

Results A total of 22 patients (age 17 -31) with DMD were reviewed (demographics summarised in table 1). All patients were evaluated with echocardiography on at least one occasion (mean EF 44.3%). Cardiac MRI was attempted in six patients, however due to contractures preventing access to the scanner only three were completed. 14 of 22 patients (64%) demonstrated an impaired left ventricular ejection fraction (EF) < 50% (mean EF 41%). Proximal ‘hot spot’ deletions/mutations (exon 2–19) appeared to be associated with a more pronounced reduction in EF – all those patients with proximal mutations demonstrated an EF < 45% (mean 41%). Seven of the eight patients (87.5%) with mutations involving >1 exon deletion demonstrated more severely impaired EF (mean EF 37.5%) compared to those with single exon deletions (mean EF 52%). Interestingly, one patient with a proximal mutation (exon 3–6 deletion) remains mobilising to distances up to 70 m, however CMRI performed has shown a moderate degree of fibrosis with an EF of 42%. Correlation between nt-Pro BNP levels and reduced EF (rEF) was not uniform, however a level < 100

Abstract 19 Table 1 Demographics

	Age	Mutation	EF	nt-Pro BNP	No. of HF Medications
I	17	Exon 3–6 Del	42%	Not Available	2
II	17	Nonsense Exon 19	45%	Not Available	2
III	18	Nonsense Exon 19	45%	Not Available	2
IV	19	Exon 47–52 Del	40%	251	3
V	20	Exon 37–43 Del	50%	300	1
VI	20	Exon 45–53 Del	30%	37	3
VII	17	Exon 44–47 Del	50%	Not Available	3
VIII	22	Exon 45–50 + 52 Del	30%	231	2
IX	21	Exon 43–53 Del	25%	37	3
X	21	Exon 45 Del	45%	290	3
XI	28	Absent on IHC	50%	35	2
XII	20	Absent on IHC	40%	164	2
XIII	31	Uncertain	20%	108	2
XIV	18	Exon 68 Del	45%	140	3
XV	20	Exon 68 Del	70%	36	1
XVI	20	Exon 45 Del	35%	290	3
XVII	19	Exon 44 Del	55%	12	2
XVIII	26	Uncertain	55%	24	1
XIX	31	Uncertain	60%	47	3
XX	17	Uncertain	55%	113	3
XXI	26	Uncertain	55%	Not Available	1
XXII	31	Exon 50 Del	60%	63	2

Nt-Pro BNP – N-terminal pro-B-type natriuretic peptide, HF – Heart Failure, IHC – immunohistochemistry, EF – ejection fraction.

was associated with EF >55% in 89% of cases. Of those on steroid regimes, 6 (54%) had impaired LVEF compared to 5 (83%) of those not on steroid therapy. All patients are taking at least one class of heart failure modification, with 79% on two and 37% on three. Only one patient in the registry has had an ICD implanted. This patient has had a device for 10 years and in this time there have been no therapies delivered.

Conclusion Correlation between predictors and cardiac phenotype in a Duchenne population remains unreliable. Location and size of exon alteration appears to be indicative of more markedly impaired LV function, however larger studies are required to characterise this further and challenges remain with regard to accurate assessment of EF. The use of predictors in future may help to guide appropriate provision of device therapy.

20 HOW DOES BMI INFLUENCE HEART FAILURE PROGRAMME OUTCOMES?-TESTING THE OBESITY PARADOX

¹L Byrne, ¹P Wheen, ²P Murray, ³C Minelli, ¹D O’Callaghan, ¹C Daly. ¹St. James’s Hospital, Dublin, Ireland; ²Tallaght University Hospital, Dublin, Ireland; ³MRHT

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Introduction A complex physiological relationship exists between obesity and heart failure (HF) with many large-scale studies reporting a paradoxical improvement in cardiovascular (CV) mortality in obese patients. Many of these studies retrospectively analyzed RCT trials not designed to investigate the role of obesity in HF outcomes. We aim to establish if