An Audit of Patients with Muscular Dystrophy Attending a UK Regional Paediatric Cardiology Department

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Objective
The aim was to examine the current practices employed by the cardiology team in the investigation, monitoring and treatment of patients with muscular dystrophy.

Methods
A search for all patients aged 0-16 years with a diagnosis of Muscular Dystrophy (MD) attending the paediatric cardiology department was performed using the Heartsuite Database. Patients who did not have a confirmed diagnosis were excluded. Electronic medical records were then accessed and examined. Details extracted included patient demographics, age at time of referral to cardiology, initial investigation findings, current cardiac status, and drug therapy. In particular, attention was paid to what measurements were being taken during echocardiogram (ECHO) assessment and the timing and choice of drug when medication was first introduced.

Results
36 patients aged between 4-16 years were identified. 77% of patients had a diagnosis of Duchenne’s Muscular Dystrophy while Becker’s Muscular Dystrophy accounted for 6%. A wide range of ages was seen at the time of initial referral to Cardiology (1-11 years). At initial assessment 94% had a normal 12 lead electrocardiogram (ECG) and 100% a normal ECHO (78% at this time had ejection fraction/shortening fraction (EF/FS) calculated while 3% had tissue doppler performed). Review intervals for patients were found to be variable ranging from 6 monthly to 3 yearly. At their most recent review, 83% had a normal ECG, while 87% had normal ECHO findings. At this stage 30% had tissue doppler performed and 60% had EF/FS calculated. Of those with abnormal ECHO findings, all had EF/FS calculated, while 75% had tissue doppler performed. Nine patients were found to be on medical therapy. Seven different drugs were prescribed including 3 different types of beta-blocker (5 patients), 3 different ACE-inhibitors (7 patients) and digoxin (1 patient). 3 patients required multi-drug therapy.

Conclusions
This audit of current practice highlighted that there was little consistency in the management of patients with MD. As a result the department has drawn up a new proposed guideline for the management of this patient group.