Non-invasive Detection of Cardiac Abnormalities in Duchenne Muscular Dystrophy

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BACKGROUND: Duchenne Muscular Dystrophy (DMD) is caused by deficiency in dystrophin protein and results in progressive skeletal muscle weakness and cardiac dysfunction. With advances in treatment of DMD, cardiac disease has become a significant cause of morbidity and mortality. Cardiac magnetic resonance imaging (CMR) has proved useful in characterizing the course of cardiac changes in DMD. For example, subtle changes, such as reductions in peak left ventricular myocardial circumferential strain (ε_{cc}), can be detected in DMD patients <10 years of age with normal ejection fraction. We hypothesized that in DMD patients the relatively increased contractility of the lateral left ventricular (LV) free wall (demonstrated previously in normal subjects) might make the region more susceptible to myocardial injury with subsequent fibrosis.
METHODS: We analyzed regional ε_{cc} from myocardial tagged CMR images using HARP™ software in 70 DMD males and 13 age-matched control males. The mid-papillary level LV slice was divided into 6 regions (anterior free wall, lateral free wall, etc) as per prior convention for coronary distribution pattern. ε_{cc} from each region was tabulated per patient. In DMD patients, myocardial delayed enhancement (MDE, a marker of myocardial fibrosis) was determined by gadolinium uptake. These results were then tabulated along with the patient's ejection fraction as well as the patient/subject's age. Patient data was subdivided based on ejection fraction and MDE status.
RESULTS: The lateral free wall regions had greater reductions in ε_{cc} ($\Delta\varepsilon_{cc}$) when compared to controls. Furthermore, MDE was consistently detected in the regions with the greatest reductions in ε_{cc} in DMD patients.
CONCLUSIONS: Changes in ε_{cc} follow a pattern showing that the regions with greatest contractility in control subjects (the lateral free wall) are the most susceptible to injury in DMD patients, as exemplified both by the greatest reduction in regional ε_{cc} and the development of MDE in those regions. Furthermore, both the ε_{cc} reduction and MDE ultimately result in global cardiac dysfunction, as there appears to be a threshold of regional strain reduction that when crossed manifest as global functional decline.