A LONGLITUDINAL EXAMINATION OF CARDIAC DYSFUNCTION IN PATIENTS WITH DUCHENNE AND BECKER MUSCULAR DYSTROPHY VIA CMRI

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Background: Duchenne and Becker muscular dystrophy (D/BMD) typically exhibit cardiac dysfunction, but the temporal evolution is not well understood. Evaluation of potential therapies for cardiac dysfunction in D/BMD patients requires accurate, quantitative, longitudinal data; therefore, our study examined the relationship of cardiac function, age, and myocardial fibrosis longitudinally in a large group of D/BMD patients.

Methods: D/BMD patients who underwent serial cardiac magnetic resonance (CMR) studies over 7.5 years were evaluated for left ventricular ejection fraction (EF) and presence of late gadolinium enhancement (LGE), a marker for myocardial fibrosis. The data were analyzed using linear mixed effects regression.

Results: A total of 336 CMR studies from 75 D/BMD patients with >=4 CMR were analyzed. Patient age at time of CMR ranged from 6.6 to 28.6 yrs (median 12.2, mean 13.1). Forty-one studies (12%) showed depressed EF (EF <55%) and 91 studies were LGE+ (27%). The youngest patient with depressed EF was 6.9 yrs and the oldest with normal EF was 28.6 yrs. The youngest LGE+ patient was 8.4 yrs and the oldest LGE− patient was 28.2 yrs. Thirty-nine patients (52%) had >=1 abnormal CMR, including 18 patients (24%) with depressed EF; 37 patients (49%) with LGE+; and 16 patients (21%) with both. Five patients (7%) were LGE+ on their first study. Twenty-six patients (35%) developed LGE before depressed EF; 11 (15%) developed depressed EF before LGE; and 2 (2.7%) developed both on the same CMR. The patients exhibited depressed EF at a median age of 14.9 yrs and LGE at a median age of 13.1 yrs. Age was associated with an EF decline of 0.57%/yr (p<0.0001) overall; however, multivariate analysis showed that EF declined 0.25%/yr when LGE is absent (p=NS) but the decline accelerated to 1.9%/yr when LGE was present (p<0.0001).

Conclusion: These data show that the temporal characteristics of cardiac function in D/BMD patients are highly variable. There is a strong association of LV dysfunction with age, but dysfunction accelerates markedly when LGE is evident. Therefore, therapeutic interventions targeting myocardial fibrosis may be beneficial in slowing the progression of cardiac dysfunction in D/BMD patients.