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Early cardiac evaluation in Duchenne or Becker muscular dystrophy using quantitative assessment of late gadolinium-enhanced magnetic resonance imaging

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Introduction: Cardiac involvement in Duchenne or Becker muscular dystrophy (DMD/BMD) has been postulated to be often overlooked. Recently, cardiac magnetic resonance imaging with late gadolinium enhancement (CMR-LGE) has enabled visualization of the fibrotic area in the myocardium, making it a reliable modality for early diagnosis of cardiomyopathy in DMD/BMD. However, quantitative assessment of CMR-LGE in DMD/BMD has not been established.

Methods: Forty-six consecutive CMR-LGE images in 26 patients with DMD/BMD (2.9-29.0 years old, median 11.0 years old) were studied retrospectively. %tLGE (ratio of LGE area to LVmass), LGE_n (number of segmental area with positive LGE), Indexed left ventricular end-diastolic volume (LVEDVI) and left ventricular ejection fraction (LVEF) were obtained at later analysis.

1) The relationship between %tLGE and LGE_n, and 2) The relationship between LVEDVI, LVEF and %tLGE, and the cut-off value of %tLGE for LVEDVI>92ml/BSA and LVEF<50% were studied. Using 26 CMR-LGE data at first scan of each patient, 3) The relationship between LVEDVI, LVEF, %tLGE and age, and the cut-off age for LVEDVI>92ml/BSA and LVEF<50% were studied. 4) The comparison of LVEDVI, LVEF, %tLGE between two groups divided by an age (Group A (n=11), <10years old, Group B (n=16), ≥10years old) were studied.

Results: 1) %tLGE were strongly correlated with LGE_n (r=0.98).

2) Both LVEDVI and LVEF were strongly correlated with %tLGE (r=0.63, 0.73 respectively). Cut-off value of %tLGE for LVEF<50% and LVEDVI>92ml/BSA were 5% and 28%, respectively.

3) With advancing years with age, %LGE increased (r=0.27), LVEF decreased (r=0.33), and LVEDVI mildly increased (r=0.42). Ten years old was a cut-off point of both LVEF<50% and LVEDVI>92ml/BSA.

4) %tLGE and LVEDVI were significantly higher in Group B. LVEF was significantly lower in Group B.

Discussion: %tLGE reflected the extension of fibrosis in LV myocardium as well as LV pump function. We concluded that 10 years old was a turning point of progressing LV fibrosis and reducing LV pump function. Therefore, cardioprotection should be considered before 10 years old.

Conclusions: Quantitative assessment of CMR-LGE could be a gold standard method for cardiac evaluation in patients with DMD/BMD.