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The relationship between clinical stage, prognosis and myocardial damage in patients with Duchenne-type muscular dystrophy: five-year follow-up study

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The evaluation of myocardial damage by [^{123}I] 15-(*p*-iodophenyl)-3-(*R,S*)-methylpentadecanoic acid (BMIPP) imaging, which represents free fatty acid metabolism, has not been reported in patients with Duchenne-type muscular dystrophy (DMD). To date, the relationship between clinical stage, prognosis and myocardial damage has not been evaluated by radionuclear cardiac imaging. The main goal of this study was to elucidate the relationship of quantitative indices of myocardial damage obtained by radionuclear cardiac imaging ([^{201}Tl] and [^{123}I] BMIPP) to clinical stage and incidence of severe cardiac events in patients with Duchenne-type muscular dystrophy (DMD). ***Methods:*** The study population consisted of 28 male patients with DMD. The average age at the beginning of observation was 19.1 ± 7.4 yrs. Nuclear tomographic imaging was performed using [^{201}Tl] and [^{123}I] BMIPP. The mid-ventricular short axial slices were classified into four anatomical regions, and the normalized count data in these areas (TL, BM) were obtained. The endpoint was the occurrence of heart failure during the follow up period. ***Results:*** Thirteen cases of heart failure occurred during the 5-year follow-up period, including three cases with cardiac death due to congestive heart failure. Clinical staging correlated directly with TL ($p = 0.0118$) and BM ($p = 0.0401$) in the whole left ventricle. In regional TL analysis, an association was observed only in the septum ($p = 0.0151$), and in the anterior ($p = 0.0361$) region. The only discrepancy between the tracer parameters (TL − BM) in the septum was observed with the radionuclear cardiac values, which exhibited a relationship with cardiac events ($p = 0.0124$). This discordance, TL < BM, was contrary to that usually observed in patients with ischemic heart disease. ***Conclusion:*** The septum is the critical area of significance for cardiac events and outcome in patients with DMD. The uptake of [^{201}Tl] in this area was representative of the clinical stage, and TL-BM correlated well with the prognosis.

Key words: cardiac event, thallium-201, iodine-123-BMIPP, radionuclide imaging, SPECT