Neuromuscular comorbidity, atrial fibrillation and left bundle branch block predict the prognosis of left ventricular hypertrabeculation/noncompaction

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Background: The prognosis of patients with left ventricular hypertrabeculation/noncompaction (LVHT) is controversially assessed. LVHT is frequently associated with neuromuscular disorders (NMDs). Aim of the study was to assess cardiac and neurological findings as predictors of mortality in LVHT-patients.

Methods: Included were patients with LVHT diagnosed between June 1995 and December 2019 in one echocardiographic laboratory. They underwent a baseline cardiologic examination and were invited for a neurological investigation. In January 2020, their survival status was assessed.

Results: LVHT was diagnosed in 310 patients (93 female, aged 53±18 years) with a prevalence of 0.4%/year. A neurologic investigation was performed in 205 patients (67%). A specific NMD was found in 33 of the investigated patients (16%), NMDs of unknown etiology in 123 (60%) and the neurological investigation was normal in 49 (24%) patients. During 86 months of follow-up, 59 patients received implanted electronic devices (cardioverter/defibrillator n=21, antibradycardic pacemakers n=11, cardiac resynchronization device/defibrillator n=22, cardiac resynchronization device n=4). During follow-up 105 patients died and 6 patients underwent heart transplantation. The mortality was 4.7%/year. By multivariate analysis, the following baseline parameters were identified as predictors of mortality: increased age (p=0.0005), inpatient-status when LVHT was diagnosed (p=0.0050), presence of a specific NMD (p=0.0187) or NMD of unknown etiology (p=0.0052), atrial fibrillation (p=0.0007) and left bundle branch block (p=0.0168).

Conclusions: LVHT patients should be systematically investigated neurologically since neurological comorbidity has a prognostic impact. Electrocardiographic abnormalities like atrial fibrillation and left bundle branch block should be considered when planning pharmacotherapy and devicetherapy. It has to be assessed by prospective studies, which measures improve the prognosis of LVHT.