Impact of neuromuscular assessment on arrhythmic risk in LMNA cardiomyopathy

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Background: The current Risk Score for Life-Threatening Ventricular Tachyarrhythmias (LTVT) in LMNA cardiomyopathy does not consider the frequent association between cardiac and neuromuscular phenotypes as a part of a multisystemic disease. In particular, the prognostic value of tendon retractions (TR) has been recently described.

Objectives: We aimed to assess the prognostic role of TR in a cohort of probands with LMNA cardiomyopathy undergoing multidisciplinary workup at a referral center.

Methods: We included 28 probands with LMNA cardiomyopathy undergoing regular, prospective follow-up at a national referral center for laminopathies. All patients underwent extensive baseline characterization, including complete neuromuscular examination and systematic assessment of TR. Indications to implantable cardioverter defibrillators (ICD) for the primary prevention of sudden cardiac death were guideline-based. Patients were clustered in tertiles according to their baseline LMNA-Risk Score. The primary endpoint was the 5-year occurrence of LTVT, namely arrhythmic cardiac death, VF, sustained VT, or appropriate ICD shocks.

Results: Of the 28 probands with LMNA cardiomyopathy (mean age 33±15 years, 50% males, mean LVEF 55±14 %), neurological assessment identified 14 patients (50%) with TR, including 8 cases with no other signs of neuromuscular involvement. At baseline assessment, the median LMNA-Risk Score was 10% (IQR 6-20%). By 5-year follow-up, 9 patients (32%) met the primary endpoint, including three patients in the lowest tertiles of risk (median LMNA-Risk Score 23, range 6-15%). While other variables showed no remarkable differences between groups, the prevalence of TR was 78% among cases experiencing LTVT and 37% otherwise. The presence of either baseline LMNA-Risk score at the highest tertile (>17%) or TR was 100% predictive of 5-year LTVT (9/9 vs. 7/19, p=0.003). Results did not change after reassessment by the end of follow-up (15±5 years).

Conclusions: Our preliminary data suggest that the inclusion of TR in risk stratification may result in a better patient selection for primary prevention ICD implant in LMNA cardiomyopathy.