Abstract: P797

Similar disease progression in probands and family members with arrhythmogenic cardiomyopathy

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Background: Arrhythmogenic cardiomyopathy (AC) is an inheritable cardiomyopathy with variable penetrance and poorly described disease progression. Probands have more severe disease than family members, but the rate of disease progression is not known.

Purpose: To compare disease progression in AC probands and family members in a longitudinal cohort study.

Methods: We included consecutive AC patients and mutation positive family members. Clinical and echocardiographic parameters were assessed according to the 2010 revised Task Force criteria (TFC) at inclusion and at last clinical follow-up. In addition, we performed strain analyses of the right ventricle (RV) and the left ventricle (LV). We assessed RV longitudinal strain averaged in 3 free-wall segments (RVLS) and LV global longitudinal strain (LVGLS). Progression rates were expressed as the annual changes in echocardiographic parameters during follow-up.

Results: We included 145 patients (47% female, 47% probands, 39±16 years old). During 6.9 (IQR: 4.6 to 9.3) years of follow-up, right ventricle outflow tract (RVOT) diameter increased from 34±7 mm to 37±7 mm (p<0.001) with an increase of 0.6±1.3 mm/year, RV fractional area change (RVFAC) decreased from 39±9 % to 36±10 % (p<0.001) with a decrease of -0.3±3.0%/year. RVLS worsened from -23.9±6.7 % to -22.7±6.6 % (p=0.04) with a worsening of 0.2±1.5%/year. LVGLS worsened from -19.2±2.8 % to -18.5±3.1 % (p=0.01) with a worsening of 0.2±0.9%/year. The increase in RVOT diameter was similar in probands (from 37±8 mm to 40±8 mm, ? 3±0 mm, p=0.001) and in family members (from 32±5 mm to 36±6 mm, ? 4±0 mm, p=0.001). Also RVFAC decreased similarly in probands (from 34±9% to 31±10%, ? 3±1 %, p=0.007) and in family members (from 44±8 % to 41±7 %, ? 3±1 %, p=0.006). There was no difference in annual progression between probands and family members in RVOT diameter (0.5±1.2 mm/year vs. 0.6±1.4 mm/year p=0.68), RVFAC (-0.3±3.8%/year vs. -0.4±2.0%/year, p=0.92), RVLS (Figure) nor in LVGLS (Figure).

Conclusion: As expected, probands had worse cardiac function and dimensions compared to family members. Importantly, progression of AC disease was similar in probands and family members, highlighting the importance of close follow up of AC family members.
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