Sex differences in disease progression and arrhythmic risk in patients with arrhythmogenic cardiomyopathy

Rootwelt-Norberg C.; Lie ØH; Chivulescu M.; Castrini AI.; Almaas V.; Lyseggen E.; Haugaa KH.; Edvardsen T.
Oslo University Hospital Rikshospitalet, Department of Cardiology, Oslo, Norway

Funding Acknowledgements: Type of funding sources: Public grant(s) – EU funding. Main funding source(s): European Research Area Network on Cardiovascular Diseases (ERA-CVD)

Background: Arrhythmogenic cardiomyopathy (AC) is an inheritable heart disease characterized by high risk of life-threatening ventricular arrhythmia. Male sex has been reported as a risk factor of high disease penetrance and arrhythmia, but data on sex-specific phenotype in AC are sparse.

Purpose: To assess sex-specific disease progression in AC patients and structural cardiac changes at time of arrhythmic event.

Methods: We included consecutive AC patients in a longitudinal cohort study. We performed echocardiography at baseline according to Task Force Criteria of 2010 with additional parameters including strain. Patients were followed with repeated echocardiographic examinations. Ventricular arrhythmia was defined as aborted cardiac arrest, sustained ventricular tachycardia or appropriate therapy by an implantable cardioverter-defibrillator. In patients with documented first time ventricular arrhythmias, echocardiographic findings obtained ±30 days around the event was noted separately.

Results: We included 191 AC patients (46% female, 51% probands, age 41 ± 17 years) of which 88% had ≥2 echocardiographic examinations during 6.9 (IQR 4.7-9.8) years of follow up. Females and males had similar progression rate of right ventricular dimensions and left ventricular function, but right ventricular function decreased more in females (Figure). Arrhythmic events occurred in 85 (45%) patients and 39 patients had an echocardiographic examination at the time of their first event. There was no difference in right ventricular diameters or right or left ventricular function between females and males at the time of first arrhythmic event (right ventricular outflow tract diameter: 35 ± 7 mm vs. 39 ± 7 mm, p = 0.16, right ventricular fractional area change: 34 ± 9 % vs. 29 ± 11 %, p = 0.26, left ventricular global longitudinal strain: -18.8 ± 3.0 % vs. 17.2 ± 2.2 %, p = 0.12, respectively).

Conclusion: Disease progression was similar in male and female AC patients indicating no accelerated disease progression in males. First arrhythmic event occurred at similar cardiac dysfunction and diameters in both sexes indicating no lower risk in females.

Abstract Figure.