

 Arrhythmias and Clinical EP**APPROPRIATE IMPLANTABLE CARDIOVERTER DEFIBRILLATOR INTERVENTIONS IN PROBANDS AND FAMILY MEMBERS WITH ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY DYSPLASIA**

Moderated Poster Contributions

Arrhythmias and Clinical EP Moderated Poster Theater, Poster Hall, Hall C

Friday, March 17, 2017, 3:45 p.m.-3:55 p.m.

Session Title: Ventricular Arrhythmias and Sudden Death Prevention

Abstract Category: 5. Arrhythmias and Clinical EP: Devices

Presentation Number: 1174M-03

Authors: *Gabriela Orgeron, Cynthia James, Crystal Tichnell, Brittney Murray, Bryana Rivers, Harikrishna Tandri, Hugh Calkins, Johns Hopkins Hospital, Baltimore, MD, USA*

Background: The arrhythmic propensity of probands and family members with ARVD/C is not the same. Clinical predictors of sustained ventricular arrhythmias in follow-up may likewise differ.

Methods: The clinical course of 318 patients from the Johns Hopkins ARVD/C Registry with 1) definite ARVD/C by 2010 Task Force Criteria (TFC) and 2) implantable cardioverter defibrillators (ICD) was analyzed. Univariate cox regression analysis identified the variables significantly associated survival from appropriate ICD therapy in follow-up.

Results: Over a median follow-up of 7 years, 68% (175/258) probands and 28% (17/60) relatives had appropriate ICD interventions. Among probands, male sex (HR: 1.56, 95% CI: 1.33-2.14; $p=0.006$) and inducible ventricular arrhythmias on electrophysiology study (HR: 2.65 CI: 1.54-4.54; $p<0.001$) were associated with ICD therapy. Symptoms at presentation (HR: 1.08 CI: 0.55-2.09; $p=0.817$), pathogenic mutations (HR: 1.32 CI: 0.97-1.80; $p=0.074$), ≥ 1000 premature ventricular contractions (PVC) on 24h holter monitor (HR: 1.51 CI: 0.83-2.74; $p=0.170$), ≥ 3 T wave inversions on electrocardiogram (HR: 1.34 CI: 0.86-2.07; $p=0.191$) and major cardiac magnetic resonance (CMR) TFC (HR: 0.98 CI: 0.62-1.54; $p=0.953$) were not significant. In contrast, among family members, ≥ 1000 PVC/24h (HR: 5.35 CI: 1.27-22.48; $p=0.022$) and major CMR TFC (HR: 5.05 CI: 1.52-16.71; $p=0.008$) were associated with ICD therapy, while sex (HR: 1.43 CI: 0.55-3.72; $p=0.452$), pathogenic mutations (HR: 2.54 CI: 0.59-10.89; $p=0.210$), symptomatic presentation (HR: 1.40 CI: 0.53-3.64; $p=0.490$), inducibility on electrophysiology study (HR: 2.02 CI: 0.62-6.51; $p=0.237$), ≥ 3 T wave inversions on electrocardiogram (HR: 3.82 CI: 0.89-16.28; $p=0.069$) were not significant.

Conclusions: Probands are more likely to require ICD therapy than their affected ARVD/C relatives (HR: 3.85, 95%CI: 2.39-6.20; $p<0.001$). While male sex and a positive electrophysiology study predict ventricular arrhythmias among probands, a high PVC burden on holter monitor and evidence of structural and functional right ventricular abnormalities predict appropriate ICD therapy in ARVD/C patients detected through family screening.