1066-27 Increasing Evidence for the Familial Nature of Arrhythmogenic Right Ventricular Cardiomyopathy

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Background: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterised by fibrofatty myocardial replacement. Complications include arrhythmia, heart failure and sudden death. A genetic basis is recognised. To assess the prevalence of familial ARVC, and the mode of inheritance we undertook prospective evaluation of ARVC families.

Method: Evaluation of 65 relatives (age range 16–47y, mean 29y, 31 male) of 26 consecutive ARVC probands (ESC/IFSC criteria) included 12 lead ECG, signal averaged ECG, Holter monitoring and echocardiography. Relatives were classified as ARVC; having cardiac abnormalities not meeting ARVC diagnostic criteria; or as normal.

Results: In 6/26 (23%) families there was >1 case of ARVC. In 7/26 (27%) families, relatives had ECG abnormalities not meeting ARVC diagnostic criteria. In total, 10/26 (38%) families either had >1 case ARVC or other cardiac abnormalities The frequency of abnormalities was higher in the ARVC families than in healthy controls ($p < 0.05$). Segregation analysis was most consistent with autosomal dominant inheritance with variable expression.

Conclusions: Our survey suggests that familial ARVC is common and the frequency of ECG abnormalities in relatives of patients with ARVC is increased. This may represent early ARVC, and suggests a broader spectrum of disease than implied by current diagnostic criteria.

1066-28 Clinical Characteristics of Patients With Arrhythmogenic Right Ventricular Cardiomyopathy and Left Ventricular Involvement

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Background: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a progressive heart muscle disease that with time leads to more diffuse right ventricular (RV) changes and left ventricular (LV) involvement, and it may culminate in heart failure.

Methods: We studied 42 patients (pts) (27 males and 15 females, mean age 29.6 ± 18 years) with clinical and/or pathological diagnosis of ARVC either at autopsy (38 pts) or heart transplantation (4 pts).

Results: Ten pts had isolated RV involvement (group A), while the other 32 (76%) had LV involvement, which was only histological in 15 (Group B), and histological and macroscopical in 17 (group C).

	Group A	Group B	Group C
Age (yrs)	20 ± 8.8	25 ± 9.7	1.39 ± 15*§
Familial history	3 (30%)	8 (53%)	6 (35%)
Disease duration	1.2 ± 2.1	3.4 ± 2.2	9.3 ± 7.3*§
Asymptomatic	7 (70%)	3 (20%)*	2 (13%)*
Syncope	3 (30%)	5 (33%)	3 (18%)
Inverted T waves V1–V4	2/2 (100%)	11/12 (92%)	14/14 (100%)
Inverted T waves V5, V6	0/2	1/12 (10%)	9/14 (71%)§
Ventricular arrhythmias	2 (20%)	11 (73%)*	14 (82%)*
Heart failure	0	0	8 (47%)*§

* = P < 0.05 vs group A; § = P < 0.05 vs group B.

Conclusions: ARVC pts with LV involvement were significantly older and more symptomatic, had a longer disease duration, more often had ventricular arrhythmias and developed heart failure.

1067 Valvular Heart Disease: Anticoagulation; Atrial Fibrillation; Drug Induced

Monday, March 30, 1998, 3:00 p.m.–5:00 p.m.
Georgia World Congress Center, West Exhibit Hall Level
Presentation Hour: 3:00 p.m.–4:00 p.m.

1067-18 Mitral Regurgitation in the Presence of Left Atrial Thrombus: A Predisposing Factor or Protective Mechanism for Embolism in Atrial Fibrillation?

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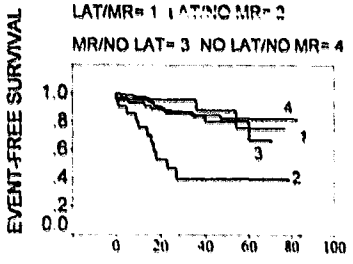
Mitral regurgitation (MR) has been proposed to decrease the incidence of embolism in atrial fibrillation (AF) by reducing stasis in the left atrium. We postulated that pre-existing left atrial thrombus (LAT) would be more likely to dislodge and embolize in the presence of MR due to increased shearing forces.

Methods: 261 pts undergoing TEE for predominantly (89%) nonvalvular AF were divided into 4 groups on the basis of LAT and significant (≥ moderate) MR: 1) Grp 1 = LAT and MR (n = 22), Grp 2 = LAT and no MR (n=24), Grp 3 = MR and no LAT (n = 62), and Grp 4 = no LAT or MR (n = 153). Pts were followed for embolism (i.e., stroke, TIA, or peripheral embolism) over a mean of 31 ± 21 months (max 85 mo).

Results: Embolism was greater in Grp 2 (50%; 12/24) vs Grp 1 (14%; 3/22), Grp 3 (15%; 9/62) or Grp 4 (12%; 19/153). Multivariate logistic regression showed that having LAT and no MR (Grp 2) was the most positive predictor of Emb (odds ratio = 6.5, p = 0.0002). Other TEE variables were not predictors. Htn was a positive predictor (odds ratio = 2.4, p = 0.05) but heart failure, and use of aspirin or warfarin were not predictors. Kaplan-Meier analysis showed a lower event-free survival in Grp 2 vs Grp 1 (p = 0.004), Grp 3 (p = 0.0007) and Grp 4 (p = 0.00001). Event-free survival did not differ among Groups 1, 3 and 4. Age did not differ. EF was lower in Grp 1 vs Grp 4 (36 ± 20 vs 51 ± 16%, p = 0.003).



Conclusions: MR in the presence or absence of LAT protects from embolism in AF. Pts with LAT and no MR are at highest risk of embolism in AF.

1067-19 The Risk of Life-Long Anticoagulation Therapy Can Be Significantly Lowered by INR Self Testing – 4-Year results of a Prospective Randomized Study

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To evaluate the influence of INR self testing (INR ST) on the risk of thrombotic, thromboembolic (TE) and bleeding complications (BL) following oral anticoagulation therapy (OAC), 150 consecutive patients (p) with the indication for life long OAC (mechanical valve replacement) were allocated to 2 random arms after a standardized training program in INR ST using the CoaguChek® monitor and self adjustment of anticoagulant dosage. 75 p (group A) were asked to measure the INR themselves every 3rd day. Anticoagulation in the remaining 75 p (group B) was managed by home physicians. During the education period all 150 p were monitored to a target therapeutic range of INR 3.5–4.0. P were asked to contact their home physicians immediately if the INR was measured ≥0.5 below/above the target range (INR corridor 3.0–4.5). All p had outpatient reexaminations every 3 months and documented all TE and BL themselves by using special documentation cards.

	A (standard management)	B (INR ST)
n (lost to follow up)	75 (2)	75 (1)
follow-up (months)	2.456	2.508
n (INR controls)	2.166	27.543
control intervals (days)	20.2 ± 7.9	3.9 ± 0.3
mean (range) INR	4.0 ± 1.5 (1.4–5.9)	3.7 ± 0.3 (2.3–4.6)
INR within corridor (%)	3.293 (53.3%)	21.047 (76.4%)
bleedings (%/year)	10.24	4.42
TE (%/year)	3.33	0.92
severe BL/TE (%/year)	2.62	0.31

In Conclusion: this randomized study with significantly ($p < 0.001$) more INR-measurements inside the target therapeutic corridor demonstrates a significant lower risk for BL/TE in p on life-long OAC performing INR ST.

MONDAY POSTER