068 THE VENTRICULAR ECTOPIC QS INTERVAL (VEQSI): DIAGNOSIS OF ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY (ARVC)

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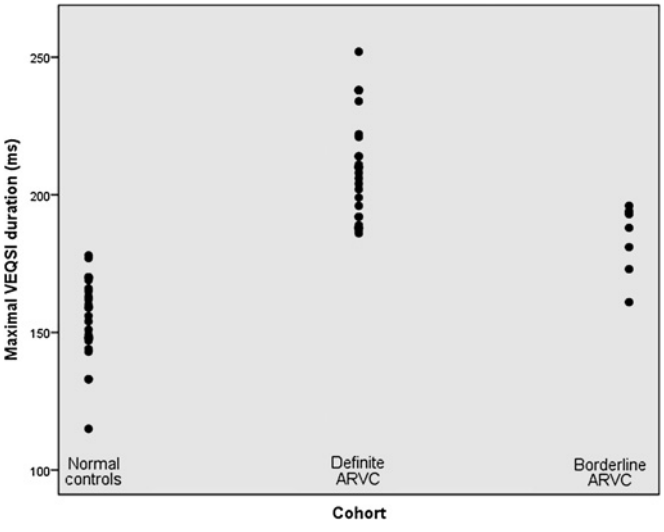
Introduction A gold standard test for arrhythmogenic right ventricular cardiomyopathy (ARVC) does not exist and diagnosis relies on meeting Task Force criteria. Sudden death remains the first clinical presentation in up to 23% of subjects. In ischaemic heart disease prolongation of the ventricular ectopic QS interval (VEQSI) has been shown to correlate with presence and severity of myocardial disease. We evaluated the significance of VEQSI in patients with ARVC.

Methods We selected 3 cohorts for 12 lead 24-h Holter monitoring: 51 normal controls (41.7±14.8 years; 55% male); 27 patients with definite ARVC by Task Force criteria (46.4±13.1 years; 70% male); 10 patients with borderline ARVC and a confirmed pathogenic mutation and/or definitely affected first degree relative (34.6±13.0 years; 50% male). All ventricular ectopics (VE) were reviewed and VEQSI was measured for each VE morphology. The longest VE duration was recorded as VEQSI max.

Results VEQSI max was significantly longer in definite ARVC compared with borderline ARVC and normal controls (208.8±18.6 ms, 183.7±12.9 ms and 155.0±14.4 ms respectively; p<0.001). VEQSI max was also significantly longer in borderline ARVC compared with normal controls (p=0.002). When the upper normal limit for VEQSI max was defined as 170 ms there was 97% sensitivity and 93% specificity for diagnosis of ARVC, including 9/10 correct diagnoses in the borderline ARVC group.

Conclusion Maximal VEQSI duration is significantly longer in definite ARVC compared with normal controls, and intermediate in ARVC patients with early disease. VEQSI max >170 ms has high sensitivity and specificity for the diagnosis of ARVC and may be particularly useful in borderline disease.



Abstract 068 Figure 1

069 SYSTEMATIC REVIEW OF 1142 ADMISSIONS WITH ACUTE HEART FAILURE REVEALS HIGH FREQUENCY OF TRANSTHYRETIN V122I CARDIAC AMYLOIDOSIS IN AFRO-CARIBBEAN PATIENTS

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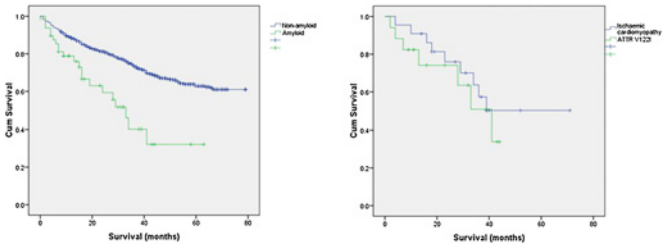
Background The genetic mutation encoding transthyretin (TTR) isoleucine 122 (V122I) is present in 3.9% of African-Americans and associated with cardiac TTR amyloidosis, but penetrance is unknown. Little is known about the frequency or clinical phenotype of TTR V122I amyloidosis in the British Afro-Caribbean population.

Methods We reviewed the primary diagnoses of 1142 patients admitted with heart failure between September 2005 and February 2011. The diagnosis was supported by echocardiography, cardiac MRI (in patients without contraindications) and genetic testing; endomyocardial biopsies were performed in 68 patients (6%) in whom amyloidosis was suspected or uncertainty about diagnosis remained. Survival analysis was performed to August 2011.

Results The median age was 72 years (range 18–98) with male (66.7%) and Caucasian (71.0%) predominance. Ischaemic cardiomyopathy (ICM) was the primary diagnosis in 428 patients (37%). There were 170 Afro-Caribbean patients (14.9%) among whom ICM was less common (22 patients (13%), p<0.01). Seventeen Afro-Caribbean patients (10%) were confirmed to have cardiac ATTR V122I amyloidosis. Survival of Afro-Caribbean patients with ICM and ATTR V122I amyloidosis was similar (45 vs 36 months, p=0.54), but overall survival (n=1142) was significantly inferior in cardiac amyloidosis compared to non-amyloid cardiomyopathy (34 vs 59 months, p<0.01).

Conclusion ATTR V122I amyloidosis is an important cause of heart failure in the British Afro-Caribbean population, but it is commonly misdiagnosed as hypertensive cardiomyopathy. Diagnosis requires specialist multidisciplinary investigation including CMR, genetic testing and histology, enabling appropriate management in an era when novel treatments for amyloidosis are entering the clinic.



Abstract 069 Figure 1 Kaplan–Meier curves for all patients (n=1142) demonstrating significantly inferior survival in cardiac amyloidosis compared to non-amyloid cardiomyopathy (p<0.01). Kaplan–Meier curves in Afro-Caribbean patients (N=170) demonstrating no significant difference in survival between ischaemic cardiomyopathy and ATTR V122I (p=0.54).