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The authors report no conflict of interest.

Word count:

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ECG in Differentiating Athlete’s Heart from Arrhythmogenic Cardiomyopathy?

Sudden cardiac death from a quiescent cardiomyopathy is a feared complication of sport in young athletes. Most elite sporting organisations in Europe implement cardiac screening with ECG in young athletes to identify affected individuals on the premise that early detection and intervention can minimise the risk of such catastrophes. ECG interpretation in young athletes is challenging because regular intensive exercise is associated several repolarisation anomalies which overlap with those detected in patients with cardiomyopathy. Early repolarisation, T wave inversion (TWI) and a prolonged QT interval are all recognised features of athletic training but are also established markers of cardiac diseases associated with high arrhythmic risk. The differentiation between cardiac electrical and structural markers of physiological adaptation and cardiomyopathy is one of the most intriguing aspects of sports cardiology and an area of evolving paradigms and research.

Anterior TWI (leads V1-V3) is the most commonly recognised electrical abnormality in individuals with arrhythmogenic right ventricular cardiomyopathy (ARVC) but is also present in approximately 10% of white endurance athletes and a larger proportion of black athletes. TWI rarely extends beyond V2 white endurance athletes (1-4%) but may affect leads V3 and V4 in 12-13% of black athletes where it is frequently preceded by J-point elevation (JPE) ≥ 0.1 mV and ST segment. Indeed, this trio of repolarisation anomalies is considered as a normal ethnic variant in black athletes and does not require further investigation in asymptomatic athletes without a relevant family history. The significance of TWI in leads V2-V4 in white endurance athletes is less clear particularly in light of emerging reports that intensive endurance exercise may result in adverse electrical and structural remodelling of the right ventricle that is indistinguishable from familial ARVC (Heidbuchel).

Early repolarisation in the anterior leads is common in athletes, therefore Calore and colleagues examined the role of preceding JPE in distinguishing athletes with anterior TWI and individuals with ARVC. Fifty seven young athletes (median age 21-years-old) with TWI in V1-V4 but no other features of ARVC were compared with 26 probands with ARVC including 9 athletes (mean age 32-years-old). The authors concluded that JPE ≤ 0.1 mV showed a 100% sensitivity for cardiomyopathy and 55% specificity for physiological adaptation. In other words, none of the patients with ARVC and TWI confined to V1-V4 showed JPE ≥0.1 mV, therefore its presence had excellent negative predictive value for ARVC. In contrast, half of the healthy athletes with anterior TWI also showed JPE ≤ 0.1 mV hence its presence provided limited clinical information.

The low specificity of JPE ≤ 0.1 mV is not surprising because the prevalence of early repolarisation in athletes does not exceed 40-50% and is most common in young black male athletes and white male endurance athletes, whereas white female athletes rarely reveal this phenomenon. The sensitivity of this parameter, when applied to the general young sporting population in the Western world, is also questionable because Calore compared predominantly black male athletes (70%) who would be expected to have a high prevalence of JPE with ARVC patients. Furthermore the number of patients in this study was relatively small to draw concrete conclusions for a conundrum where an erroneous diagnosis could have potentially serious implications.

 In this issue of JACC Clinical Electrophysiology, Brosnan and colleagues revisit electrocardiographic features that facilitate the differentiation between athlete’s heart and ARVC. Electrocardiograms in 100 athletes with anterior TWI (62% in V1 and V2 only) were compared with ECGs in 100 patients with ARVC who were matched for age, sex and ethnicity. Both cohorts had a mean age between 21.5 and 22.5 years respectively, 97% were white and 53% were female. The ARVC patients were recruited from the Johns Hopkins ARVD/C Registry and The Netherlands Heart Institute ARVC Registry. In contrast with Calore and colleagues, the investigators did not notice any difference in the prevalence of JPE ≥ 0.1 mV between athletes and patients with ARVC (27% v 16%; p = 0.09). The prevalence of JPE ≥ 0.1 mV in patients with ARVC is much higher than previous reports in patients who were between 10-20 years older than the current cohort (Calore and Finocchiaro) and could be due to several age related factors including progression of disease severity and changes in cardiac vagal tone. JPE ≤ 0.1 mV preceding anterior TWI (V1-V4) showed a sensitivity of 92% for detecting ARVC this young, non-black and predominantly female cohort. Although this parameter would not have detected 8% of affected individuals, this figure is not dissimilar to that reported for a normal electrocardiogram in young individuals with hypertrophic cardiomyopathy. Of more interest was the 10-fold higher prevalence of JPE ≥0.2mV in athletes compared with patients with ARVC suggesting that higher JPE is more useful for excluding ARVC with a sensitivity reaching 98%. The specificity of JPE for predicting athlete’s heart is even poorer in white athletes and should not be considered further.

The authors identified several other parameters that were more predictive of ARVC than athlete’s heart including T wave inversion beyond V3 or inferior leads, premature ventricular complexes (PVC) on a 10 second ECG and low limb lead voltages. T wave inversion beyond V4 and PVCs were not identified in any athlete Furthermore, T wave inversion in the inferior leads identified in only 3% athletes compared with 31% patients with ARVC and low limb lead complexes were noted in only 1 athletes compared with 21% of ARVC patients. A separate analysis in 30 ARVC patients who exercised regularly also revealed a much higher prevalence of these anomalies compared with healthy athletes.

The strength of this study is the matched design for similar age, sex and ethnicity between ARVC patients and athletes, all of which are confounders in prevalence of JPE. Apart from a lower sensitivity of JPE ≤ 0.1 mV for detecting ARVC in this cohort, the findings are not dissimilar to those reported recently by Finocchiaro and colleagues who investigated 129 healthy individuals (athletes) of similar age who were compared with 82 patients with ARVC who were 20 years older. Healthy individuals had been investigated comprehensively for ARVC. Although only 2% of this older cohort of ARVC patients showed JPE ≥ 0.1 mV, these patients had a higher prevalence of other ECG anomalies in addition to anterior TWI (77% v 19%). The authors excluded young healthy individuals with inferior and lateral TWI because these anomalies are considered abnormal and warrant in white athletes and warrant investigation. As with this study over 30% of ARVC patients had inferior TWI and 29% showed lateral TWI. The prevalence of lower limb lead complexes was higher in ARVC patients (15% v 4%). In contrast PVCs were observed in 2% of healthy and this may be reflected by more advanced disease. Additionally 28% ARVC patients showed S wave duration in V2 > 55 msec compared to none of the healthy athletes.

There have been several developments in our understanding of the ECG in patients with ARVC. Whereas anterior TWI in V1-V3 to patients with ARVC and some athletes, there are several other anomalies that appear to be strong discriminators between the two entities. The heterogeneous nature of ARVC, variable involvement of the left ventricle and overt contribution of environmental stimuli to disease phenotype represents a challenge in finding single parameters with high sensitivity and specificity, however an algorithm that incorporates a plethora of anomalies is beginning to pave an optimistic path.

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