



Original Article

Evaluating the 12-Lead Electrocardiogram for Diagnosing ARVC in Young Populations: Implications for Preparticipation Screening of Athletes

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ABSTRACT

Background: Arrhythmogenic right-ventricular cardiomyopathy (ARVC) is an identified cause of sport-related sudden cardiac arrest (SCA). Identifying athletes with ARVC and restricting them from exercise is believed to reduce the risk of SCA. The electrocardiogram (ECG) is considered to be an important component of screening for ARVC; however, the sensitivity of the 12-lead ECG to identify ARVC in young asymptomatic persons is unknown.

Methods: In this retrospective study, we identified 70 patients (49 ARVC-positive, based on Task Force Criteria, and 21 age-matched ARVC-negative persons from a paediatric arrhythmia database (<18 years of age); ECGs were analyzed for abnormalities, based on International Criteria for Interpretation of ECGs in Athletes, and ECG findings were adjudicated by group consensus.

Results: Of the 49 ARVC-positive patients (median age: 17 [interquartile range: 16-18], 65% male), 22% were found to have abnormal ECGs; the most common ECG findings were T-wave inversions. Patients

RÉSUMÉ

Introduction : La cardiomyopathie ventriculaire droite arythmogène (CVDA) est une cause établie d'arrêt cardiaque soudain (ACS) du sportif. On croit que le fait de repérer les athlètes atteints de CVDA et de limiter leur exercice réduit le risque d'ACS. On considère que l'électrocardiogramme (ECG) est une composante importante du dépistage de la CVDA. Toutefois, on ignore la sensibilité de l'ECG à 12 dérivations pour détecter la CVDA chez les jeunes personnes asymptomatiques.

Méthodes : Dans cette étude rétrospective, nous avons repéré 70 patients (49 personnes dont les résultats sont positifs à la CVDA selon les critères de la Task Force, et 21 personnes appariées selon l'âge dont les résultats étaient positifs à la CVDA) d'une base de données sur l'arythmie en pédiatrie (< 18 ans); nous avons analysé les ECG pour détecter les anomalies selon les Critères de consensus internationaux de l'interprétation de l'ECG chez l'athlète, et nous sommes prononcés sur les résultats des ECG par consensus.

Arrhythmogenic right-ventricular cardiomyopathy (ARVC) is an inherited form of structural heart disease that predisposes individuals to ventricular arrhythmias and sudden cardiac arrest (SCA) and is an important cause of sport-related SCA.^{1,2} Most people who develop symptoms present between the

second and fifth decades of life with palpitations, syncope, or SCA.^{3,4} The age at which symptoms appear is variable, but most patients eventually become symptomatic.⁵ More intense and more prolonged exercise is associated with earlier presentation and more severe phenotype.⁶ It is believed that if persons with ARVC can be identified at a young age, before participation in strenuous exercise or competitive sports, their disease progression can be modified and their risk of SCA reduced, if they are restricted from sports.⁵

Preparticipation screening programs of young athletes, particularly with respect to screening for ARVC, are designed to identify persons at risk of SCA and implement preventive measures (such as sports restriction, disease-modifying therapy, or prevention with implantable defibrillators). All preparticipation screening programs worldwide recommend initial

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with symptoms were more likely to have abnormal ECGs than asymptomatic patients (28% compared with 17%, respectively; $P = 0.002$). Of 16 gene-positive patients, 31% had abnormal ECGs. Patients with abnormal ECGs had larger right-ventricular end-diastolic volume indexes on magnetic resonance imaging than those with normal ECGs ($P = 0.03$).

Conclusions: The ECG was insensitive for detecting ARVC in young (age <18 years), asymptomatic patients, and is unlikely to provide significant diagnostic value for identifying ARVC on routine pre-participation screening of adolescent athletes.

evaluation with history and physical examination, and most also recommend a 12-lead electrocardiogram (ECG); if these test results are all normal, no guideline recommends further investigations with more advanced testing.⁷ The potential identification of ARVC has often been cited as a reason to include the 12-lead ECG in initial screening assessments, as it is reported to be > 80% sensitive.⁸ However, the utility of 12-lead screening ECG as a component of preparticipation evaluation is influenced by the sensitivity of the ECG to detect disease in this specific setting. The sensitivity data usually quoted derives from studies of older individuals (with potentially more advanced ARVC phenotypes), as opposed to adolescents (with potentially milder ARVC phenotypes), a typical age for preparticipation screening. Furthermore, it remains uncertain if preparticipation screening has a significant impact on the incidence of SCA. In a recent study of SCA in young (age < 45) competitive athletes in Toronto, Canada, where sports restriction is not practiced, ARVC was not found to be a cause of SCA in any of the 16 athletes identified over the 6-year study period.⁹ We therefore examined the sensitivity of the ECG to detect abnormalities associated with known ARVC in young persons (age < 18 years), and particularly in those without symptoms, to ascertain the potential sensitivity of a screening ECG for ARVC in this population.

Methods

Study design

In this retrospective study, we identified 49 patients aged < 18 with ARVC (definite or borderline) from a pediatric arrhythmic clinic database at The Hospital for Sick Children in Toronto, Canada. An additional 21 age-matched (< 18 years) ARVC-negative individuals were also identified (most of whom were relatives of ARVC-positive patients and were confirmed phenotype and/or genotype negative, based on imaging studies and genetic analysis) to verify the accuracy of ECG interpretation.

Authors (C.H.L, K.A.C., P.A., and P.D.) reviewed the most recent ECGs available in the database for all 70 study participants, blinded to patient diagnosis (patient age was available for

Résultats : Parmi les 49 patients dont les résultats étaient positifs à la CVDA (âge médian : 17 [intervalle interquartile : 16-18], 65 % de sexe masculin), nous avons observé que 22 % d'entre eux avaient des résultats anormaux à l'ECG; les résultats les plus fréquents à l'ECG montraient des inversions de l'onde T. Les patients symptomatiques étaient plus susceptibles d'avoir des résultats anormaux à l'ECG que les patients asymptomatiques (28 % et 17 %, respectivement; $P = 0,002$). Parmi les 16 patients qui avaient des résultats génétiques positifs, 31 % avaient des résultats anormaux à l'ECG. Les patients qui avaient des résultats anormaux à l'ECG avaient des indices plus grands du volume télédiastolique du ventricule droit en fin de diastole à l'imagerie par résonance magnétique que ceux qui avaient des résultats normaux à l'ECG ($P = 0,03$).

Conclusions : L'ECG n'a pas montré la sensibilité pour détecter la CVDA chez les jeunes (< 18 ans) patients asymptomatiques et n'a pas été susceptible de fournir une valeur diagnostique significative pour détecter la CVDA au dépistage systématique préalable à l'activité sportive des athlètes adolescents.

each ECG). ECGs were recorded as normal or abnormal according to the International Criteria/Recommendations for Electrocardiographic Interpretation in Athletes,^{10,11} using definitions for abnormal T-wave inversions as detailed by Finocchiaro et al.⁶ ECG interpretation was conducted by 2 adult cardiac electrophysiologists, a sports cardiologist expert in cardiac imaging, and a medical resident with training in sports cardiology (combined ECG reading experience of approximately 62 years). ECG results were printed and read.

Specific abnormalities coded included axis, T-wave inversions, ST changes, Q waves, bundle branch blocks, Δ waves/Wolff-Parkinson-White, ϵ waves, ST changes consistent with Brugada phenotype, prolonged QT, profound sinus bradycardia (< 30 beats per minute), atrioventricular (AV) block, atrial tachyarrhythmias, ventricular arrhythmias, premature ventricular contractions (PVCs), and terminal activation duration of QRS ≥ 55 milliseconds (msec).² If there were disagreements among reviewers (ie, if any reviewer's ECG coding differed from any of the other reviewers, including overall coding of abnormal vs normal, as well as any specific abnormalities recorded), interpretations were rereviewed (again blinded to diagnosis), and a final diagnosis was made by group consensus (K.A.C., P.D., and P.A.).

Case identification/database

The ARVC database at The Hospital for Sick Children includes patients < 18 years old who were identified from patients referred for symptoms or documented ventricular arrhythmias or via cascade screening of gene-positive family members. This cohort of adolescents is not necessarily composed of athletes at the time of entry into the database.

All diagnoses of ARVC were based on 2010 Revised Task Force Criteria¹² and included data from 12-lead ECGs, signal-averaged ECGs (SAECGs), echocardiograms (ECHOs), cardiac magnetic resonance imaging (cMRI), and genetic analyses. Specific imaging details and parameters, as well as genetic analysis testing, have been described previously.¹³⁻¹⁵ Based on the Revised Task Force Criteria, a definite diagnosis includes 2 major criteria, or 1 major criterion and 2 minor criteria, or 4 minor criteria from different categories; borderline diagnosis includes 1

Table 1. Details of investigations

Investigations	All ARVC* (n = 49)	ARVC (n = 49)	
		With symptoms (n = 25)	Without symptoms (n = 24)
Abnormal ECG [†]	11 (22%) [‡]	7 (28%)	4 (17%)
Abnormal SAECG	29 (59%)	14 (56%)	15 (63%)
MRI major criteria [§]	27 (55%)	16 (64%)	11 (46%)
MRI minor criteria	8 (16%)	2 (8%)	6 (25%)
Gene positive	16 (33%)	7 (28%)	9 (38%)

Detailed MRI data was available for 38 of 49 (78%) patients. Of the 11 patients without information, 3 of 11 MRIs were not completed; 3 of 11 MRI reports that did not include detailed numerical data indicated overall normal MRIs; 5 of 11 MRI reports that did not include detailed numerical data reported findings suggestive of ARVC.

ARVC, arrhythmogenic right-ventricular cardiomyopathy; ECG, electrocardiogram; MRI, magnetic resonance imaging; RV, right ventricle; RVEDVI, right-ventricular end-diastolic volume index; SAECG, signal-averaged ECG.

* Including definite and borderline ARVC.

[†] Fleiss' $\kappa = 0.69$.

[‡] Proportional 95% confidence interval (lower bound 0.107; upper bound 0.341).

[§] Major MRI criteria: regional RV akinesia, dyskinesia or dyssynchronous RV contraction and ratio of RVEDVI ≥ 110 mL/m² (male)/ ≥ 100 mL/m² (female), or RV ejection fraction $\leq 40\%$.

^{||} Minor MRI criteria: regional RV akinesia, dyskinesia or dyssynchronous RV contraction and ratio of RVEDVI ≥ 100 to < 110 mL/m² (male)/ ≥ 90 to < 100 mL/m² (female), or RV ejection fraction $> 40\%$ to $\leq 45\%$.

major and 1 minor criterion, or 3 minor criteria from different categories; possible diagnosis includes 1 major criterion or 2 minor criteria from different categories.

Statistical analysis

Descriptive statistics were used to assess the distribution of variables; continuous variables were summarized as mean values with standard deviations, and categorical variables were summarized as counts and percentages. All calculations and analyses were performed with the use of SPSS software, version 23.0 (SPSS Software, IBM, Chicago, IL). Statistical analyses included χ^2 tests, Mann-Whitney U tests, and Pearson correlation where appropriate. A *P* value < 0.05 was considered significant.

Table 2. Clinical characteristics of ARVC-positive patients with a normal vs abnormal 12-lead ECG

	Normal 12-lead ECG (n = 38)	Abnormal 12-lead ECG (n = 11)
Age (years)	15.9 \pm 3.4	16.5 \pm 1.7
Sex (male/female)	26 male; 12 female	6 male; 5 female
Average MRI RVEDVI Size (mL/m ²)	114.9 \pm 20.3 mL/m ² *	166.2 \pm 73.8 mL/m ² [†]
Average filtered SAECG QRS duration (ms)	115.8 \pm 14.7 ms	120.2 \pm 27.8 ms
Gene positive for a pathogenic ARVC Mutations (%)	28.9 % (11 of 38)	45.5% (5 of 11)
Symptomatic (%)	45% (17 of 38)	64% (7 of 11)

ARVC, arrhythmogenic right-ventricular cardiomyopathy; ECG, electrocardiogram; MRI, magnetic resonance imaging; RVEDVI, right-ventricular end-diastolic volume index; SAECG, signal averaged ECG.

* 29 of 38 MRI data points available.

[†] 9 of 11 MRI data points available.

Results

Study participants

There were 49 patients with either definite (n = 39) or borderline (n = 10) ARVC, 32 male patients and 17 female patients, with a median age of 17 (interquartile range [IQR]: 16-17). There were 25 patients with symptoms and 24 asymptomatic patients at the time of entry into the database (5 patients with family histories of ARVC developed symptoms over time, and the index ECG was the latest ECG at the time they had become symptomatic). Of patients identified through symptoms, 15 had presyncope or syncope, 5 had sustained ventricular tachycardia, and 5 had palpitations. Of patients without symptoms, 19 were referred for positive family histories, and 5 were referred for incidental findings on physical examination or ECG.

Among the 21 persons who were phenotype (+genotype) negative for ARVC and served as ECG controls, there were 8 male subjects and 13 female subjects, with a median age of 15 (IQR: 13-16); 16 individuals had been referred for positive family history of ARVC, and 5 had no cardiac disease and no family history and were referred for other reasons.

Details of investigations and ECG findings for ARVC patients

Investigation results are outlined in Table 1. Among patients with ARVC, 59% had abnormal SAECGs, 71% had abnormal MRIs (major or minor criteria), and 33% were gene positive. Detailed numerical MRI right-ventricular (RV) volume data were available for 38 of 49 (78%) patients. A complete listing of ARVC criteria for all subjects is provided in Supplemental Table S1.

In the blinded ECG interpretation, we identified 22% of the ECGs as abnormal. Clinical characteristics of patients with normal vs abnormal ECGs are depicted in Table 2, and specific details of ECG abnormalities are outlined in Table 3. None of the ECGs determined to be normal had ECG abnormalities that would have prompted further investigations in the context of preparticipation screening. All 21 ECG control subjects had normal ECGs with no abnormalities identified on blinded reading in any of these subjects.

Patients with symptoms were more likely to have abnormal ECGs (28% compared with 17% in asymptomatic patients; *P* = 0.002). The average age of patients with abnormal ECGs was 16.5 years, similar to the overall average age in the database of 16 years. The most common ECG abnormality found was abnormal T-wave inversions in both the symptomatic (20%) and asymptomatic (13%) cohort. None of the patients had an ϵ wave. An abnormal ECG was more likely to be found in patients with diagnoses of definite ARVC (10 of 11 abnormal ECGs) compared with those who had diagnoses of borderline ARVC (1 of 11 abnormal ECGs). Of 16 patients who were gene positive, 5 had abnormal ECGs (31%); 4 patients had pathologic gene mutation in the desmosomal protein plakophilin-2 (PKP2); and 1 had pathologic mutation in a cardiac sodium channel (SCN5A gene).

There was ECG reader disagreement in 9 of 70 (13%) patient ECGs, in which at least 1 reader coded at least 1 ECG variable as "normal" or "abnormal" differently from the other readers. The main discrepancies were in coding the axis as abnormal (in 3 of 9), when upon review consensus found

Table 3. ECG findings in patients with ARVC

ECG finding	Definite/borderline ARVC (n = 49)*	
	With symptoms (n = 25)	Without symptoms† (n = 24)
Any abnormality	7 (28%)	4 (17%)
Abnormal T-wave inversion‡	5 (20%)	3 (13%)
> 1 premature ventricular complex	2 (8%)	2 (8%)
ε waves	0	0
Borderline criteria (includes right axis deviation and complete RBBB)	1 (4%)	1 (4%)
Terminal activation duration of the QRS (> 55 ms)	1 (4%)	1 (4%)

ARVC, arrhythmogenic right-ventricular cardiomyopathy, ECG, electrocardiogram; RBBB, right bundle branch block.

* Definite ARVC, n = 39 with 10 of 11 abnormal ECGs; borderline ARVC, n = 10, with 1 of 11 abnormal ECGs (> 1 PVCs).

† Includes referrals because of positive family history of ARVC or incidental findings.

‡ T-wave inversions were anterior (5 of 8), anterolateral (2 of 8), and biphasic (1 of 8).

them to be normal and coding T-wave changes as normal (in 3 of 9), when consensus review found them to be abnormal. The inter-rater reliability as calculated by Fleiss' κ was 0.69.

Patients with abnormal ECGs had larger RV end-diastolic volume index (RVEDVI) on MRI compared with those who had normal ECGs (Fig. 1; $P = 0.03$). There was a significant, moderate positive correlation between filtered QRS duration on the SAECG and RVEDVI ($r = 0.621$, $P < 0.001$).

Discussion

The main finding in this study is that the 12-lead ECG is not sensitive in the detection of ARVC in adolescents (using the International Criteria/Recommendations for Electrocardiographic Interpretation in Athletes), with any ECG abnormality detected in only 22% of patients with confirmed ARVC as per Task Force Criteria. This is in contrast to previous reports that have suggested that the 12-lead ECG

is > 80% sensitive in detecting ARVC.¹¹ The European Sports Recommendations explicitly state that the ECG is abnormal in > 60% of persons with ARVC.¹⁶

The low sensitivity of the ECG in the diagnosis of ARVC in adolescents in our cohort has important clinical implications for incorporating the ECG into routine preparticipation screening of young athletes. Early detection of ARVC and subsequent restriction from sports has been recommended as a reasonable therapeutic intervention (ie, as exercise worsens the ARVC phenotype and is associated with a higher risk of arrhythmias, restriction of sports may be therapeutic).⁵ This intervention, however, may have a negligible effect on the overall incidence of SCA in young athletes. Notably, ARVC was not found to be a cause of SCA in any of the young competitive athletes with SCA over a 6-year period in Toronto, Canada, where sports restriction is not practiced.⁹

The utility of screening for disease detection depends on the sensitivity of the screening test; previous studies, which report > 80% sensitivity only assessed the sensitivity of ECG screening in older persons (mean age 41 ± 12 years) identified based on symptoms, in whom disease pathology is more likely to be advanced and possibly more easily detectable with an ECG.¹⁷⁻¹⁹ To our knowledge, this is the first study that has examined the sensitivity of the ECG to detect ARVC abnormalities in young patients < 18 years of age, a typical age range in which preparticipation screening programs would be first implemented,²⁰ and, importantly, also examined asymptomatic persons. An important distinction between this and previous studies is that this study aimed to test the potential sensitivity of the ECG in a screening paradigm of asymptomatic individuals, as it would be usually applied in the real-world setting; this is in contrast to a case-finding scenario, in which any suspicion of ARVC (based on history, physical, symptoms, etc) would result in extensive and targeted investigation. Moreover, our study population represents a best-case scenario for the detection of ARVC with respect to sensitivity and specificity. Thus, using conventionally employed international ECG criteria in the context of preparticipation screening ECGs for asymptomatic adolescent athletes may result in even lower sensitivity and specificity than observed in our study.

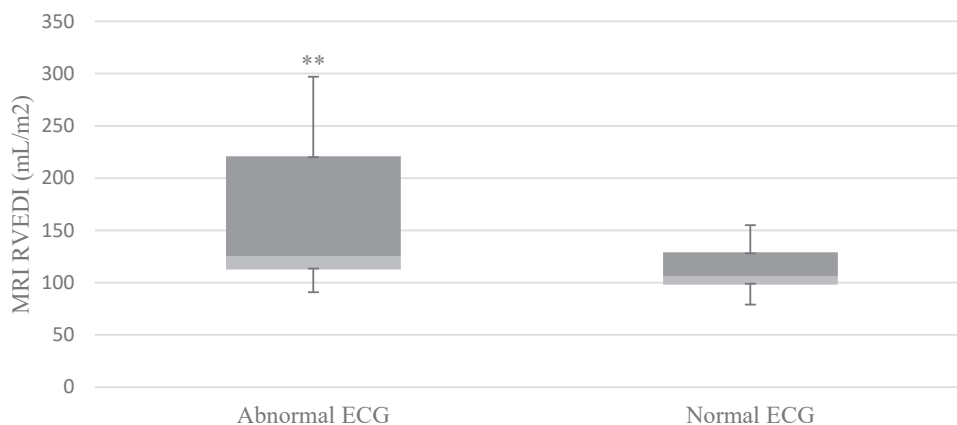


Figure 1. Abnormal vs normal ECGs and MRI RVEDVI mL/m². ECG Abnormal: any ECG abnormality present. ** $P = 0.03$, Mann-Whitney Test. Box plots represent median, interquartile range (IQR), range. Abnormal ECG RVEDVI 95% confidence interval (CI), 109.4-222.9. Normal ECG RVEDVI 95% CI, 102.5-118.7. ECG, electrocardiogram; MRI, magnetic resonance imaging; RVEDVI, right ventricular end-diastolic volume index.

The most common ECG abnormalities associated with ARVC in this study were T-wave inversions, in keeping with other studies.^{11,21} There was a modest difference in ECG sensitivity between asymptomatic and symptomatic patients (17% vs 28%, respectively); only 1 of 4 asymptomatic patients did not have any family history and therefore could only have been identified through blanket ECG screening. Patients with abnormal ECGs were also found to be more likely to have larger RV dimensions (RVEDVi). This finding may help explain the observed lower sensitivity of the ECG in this study of paediatric patients compared with that previously reported in adults, suggesting that early disease (ie, disease in younger patients with likely more mild pathology) may be less likely to be associated with ECG abnormalities. We would expect the percentage of abnormal ECGs to increase with age, especially in athletes, as the RV remodels under stress and over time.

The observed low sensitivity of the ECG to identify ARVC in young asymptomatic patients is further confounded by low ECG specificity. Low specificity makes distinguishing between an athlete's heart and ARVC on ECG alone extremely challenging owing to the high frequency of ECG changes associated with normal physiological adaptations (eg, RV enlargement) in the athlete's heart.²²⁻²⁴ In athletes with anterior T-wave inversions (TWIs), the combination of J-point elevation ≥ 1 mm and anterior TWI not extending beyond V4 excluded a cardiomyopathy, either ARVC or HCM, with 100% sensitivity and 55% specificity.¹⁷

Other ECG criteria have also not been proven to be more helpful than TWIs. For example, right bundle branch block is a nonspecific finding, and ϵ waves are insensitive and often ECG-reader dependent. The terminal activation duration of the QRS (being abnormal if ≥ 55 msec) has been reported to be a sensitive and specific ECG marker for ARVC; however, this is challenging in practice to measure and impractical as part of a mass ECG-screening process with standard 25 mm/sec paper speed recordings. The SAECG, which magnifies the QRS complex by autocorrelating and summing at least 250 beats, is likely a more useful tool, and in this study results were abnormal in 63% of asymptomatic patients with ARVC; however, it also has practical limitations and is not recommended in any current screening guideline.

At present, the European Society of Cardiology (ESC) advocates that preparticipation screening include routine 12-lead ECG screening in addition to standard history and physical examination.²⁵

The American Heart Association (AHA) guidelines do not recommend routine ECG screening; however, they also do not oppose ECG-based screening initiatives in some communities.^{26,27} In Canada, 2018 guidelines also recommend history and physical but do not recommend routine ECG screening.⁷ All guidelines recommend added tests if there are findings on history or physical examination; therefore, the sensitivity of blanket ECG screening is most relevant in asymptomatic persons. Our findings suggest that the 12-lead ECG has very low sensitivity in this cohort.

Study limitations

Limitations of this study include a small sample size ($n = 49$ patients with ARVC) and the fact that patients in the ARVC database were not necessarily athletes at the time of

entry. Although our sample size is smaller than other larger studies that report ECG sensitivity as $>80\%$,^{8,12} this is the first study to report data on young persons at the age when first screening is usually performed. Adolescent athletes who do not have ARVC would have been an ideal control group; however, our objective was to assess ECG sensitivity and not specificity. The ECGs that were analyzed were also the most recent in the database, as opposed to ECGs at the time of diagnosis. It is expected, however, that ECG results would be more likely to become abnormal as patients age, and thus using the latest ECG—when the patients were older—could potentially increase the ECG sensitivity. Our sample from Ontario, Canada may also not be representative of other regions with a higher incidence and possibly different phenotypic profile of ARVC (eg, Newfoundland or Italy).²⁸ The database did not include racial information, and thus potential differences could not be analyzed. Variability in ECG interpretation is another limitation of any study examining ECG sensitivity, as has been described,²⁹ and may influence the reproducibility of the sensitivity values we have calculated (our κ was slightly higher compared with previous reports among expert cardiologist of $\kappa = 0.40$ to 0.53); this is also a limitation of real-world preparticipation screening. More subtle ECG findings of ARVC were not analyzed, which may have decreased ECG sensitivity;^{8,30} however, these are typically found in adults with more established disease, and these criteria are not used or recommended in routine athlete screening and thus not applicable to our study objectives (or applicable to real-world athlete screening). Finally, all patients in this study either had symptoms suggesting arrhythmia disorder, relevant family history, or abnormal laboratory tests including abnormal results of ECGs. As ECGs were also used in diagnosis, the study population is not a screening population of asymptomatic persons, and the results are biased toward a higher proportion of abnormal ECGs than would be expected in an asymptomatic population with no relevant family or personal history of screening before sports preparticipation.

Conclusions

The diagnosis of ARVC in the asymptomatic young athlete remains an extraordinary challenge, and our results suggest that the addition of a routine 12-lead ECG to screening is unlikely to provide important benefit over history and physical for identifying this structural heart disease.

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Disclosures

The authors have no conflicts of interest to disclose.

References

1. Calkins H, Corrado D, Marcus F. Risk stratification in arrhythmogenic right ventricular cardiomyopathy. *Circulation* 2017;136:2068-82.
2. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. *Circulation* 2010;121:1533-41.

3. Corrado D, Basso C, Judge DP. Arrhythmogenic cardiomyopathy. *Circ Res* 2017;121:784-802.
4. Corrado D, Link MS, Calkins H. Arrhythmogenic right ventricular cardiomyopathy. *N Engl J Med* 2017;376:61-72.
5. Towbin JA, McKenna WJ, Abrams DJ, et al. 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. *Heart Rhythm* 2019;16:e301-72.
6. Lie ØH, Dejaard LA, Saberniak J, et al. Harmful effects of exercise intensity and exercise duration in patients with arrhythmogenic cardiomyopathy. *JACC Clin Electrophysiol* 2018;4.
7. Johri AM, Poirier P, Dorian P, et al. Canadian Cardiovascular Society/Canadian Heart Rhythm Society joint position statement on the cardiovascular screening of competitive athletes. *Can J Cardiol* 2019;35:1-11.
8. Finocchiaro G, Papadakis M, Dhutia H, et al. Electrocardiographic differentiation between 'benign T-wave inversion' and arrhythmogenic right ventricular cardiomyopathy. *Europace* 2019;21:332-8.
9. Landry CH, Allan KS, Connelly KA, Cunningham K, Morrison LJ, Dorian P. Sudden cardiac arrest during participation in competitive sports. *N Engl J Med* 2017;377:1943-53.
10. Drezner JA, Sharma S, Baggish A, et al. International criteria for electrocardiographic interpretation in athletes: consensus statement. *Br J Sports Med* 2017;51:704-31.
11. Sharma S, Drezner JA, Baggish A, et al. International recommendations for electrocardiographic interpretation in athletes. *J Am Coll Cardiol* 2017;69.
12. Corrado D, Wichter T, Link MS, et al. Treatment of arrhythmogenic right ventricular cardiomyopathy/dysplasia: an international task force consensus statement. *Eur Heart J* 2015;36:3227-37.
13. Etoom Y, Govindapillai S, Hamilton R, et al. Importance of CMR within the task force criteria for the diagnosis of ARVC in children and adolescents. *J Am Coll Cardiol* 2015;65:987-95.
14. Chungsomprasong P, Hamilton R, Lunning W. Left ventricular function in children and adolescents with arrhythmogenic right ventricular cardiomyopathy. *Am J Cardiol* 2016;119:778-84.
15. Chatterjee D, Fatah M, Akdis D, et al. An autoantibody identifies arrhythmogenic right ventricular cardiomyopathy and participates in its pathogenesis. *Eur Heart J* 2018;39:3932-44.
16. Pelliccia A, Solberg EE, Papadakis M, et al. Recommendations for participation in competitive and leisure time sport in athletes with cardiomyopathies, myocarditis, and pericarditis: position statement of the Sport Cardiology Section of the European Association of Preventive Cardiology (EAPC). *Eur Heart J* 2019;40:19-33.
17. Calore C, Zorzi A, Sheikh N, et al. Electrocardiographic anterior T-wave inversion in athletes of different ethnicities: differential diagnosis between athlete's heart and cardiomyopathy. *Eur Heart J* 2016;37:2515-27.
18. Cox MG, Nelen MR, Wilde AAM, et al. Activation delay and VT parameters in arrhythmogenic right ventricular dysplasia/cardiomyopathy: toward improvement of diagnostic ECG criteria. *J Cardiovasc Electrophysiol* 2008;19:775-81.
19. Steriotis AK, Bauce B, Daliento L, et al. Electrocardiographic pattern in arrhythmogenic right ventricular cardiomyopathy. *Am J Cardiol* 2009;103:1302-8.
20. Malhotra A, Dhutia H, Finocchiaro G, et al. Outcomes of cardiac screening in adolescent soccer players. *N Engl J Med* 2018;379:524-34.
21. Zaidi A, Sheikh N, Jongman JK, et al. Clinical differentiation between physiological remodeling and arrhythmogenic right ventricular cardiomyopathy in athletes with marked electrocardiographic repolarization anomalies. *J Am Coll Cardiol* 2015;65:2702-11.
22. Maron BJ, Gardin JM, Flack JM, Gidding SS, Kurosaki TT, Bild DE. Prevalence of hypertrophic cardiomyopathy in a general population of young adults: echocardiographic analysis of 4111 subjects in the CARDIA study: coronary artery risk development in (young) adults. *Circulation* 1995;92:785-9.
23. Montgomery JV, Harris KM, Casey SA, Zenovich AG, Maron BJ. Relation of electrocardiographic patterns to phenotype expression and clinical outcome in hypertrophic cardiomyopathy. *Am J Cardiol* 2005;96:270-5.
24. Bosman LP, Cadrin-Tourigny J, Boufiss M, et al. Diagnosing arrhythmogenic right ventricular cardiomyopathy by 2010 Task Force Criteria: clinical performance and simplified practical implementation. *EP Europace* 2020;22:787-96.
25. Corrado D, Pelliccia A, Bjornstad HH, et al. Cardiovascular preparticipation screening of young competitive athletes for prevention of sudden death: proposal for a common European protocol. Consensus Statement of the Study Group of Sport Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology. *Eur Heart J* 2005;26:516-24.
26. Maron BJ, Thompson PD, Ackerman MJ, et al. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update: a scientific statement from the American Heart Association Council on Nutrition, Physical Activity, and Metabolism. *Circulation* 2007;115:1643-55.
27. Maron BJ, Levine BD, Washington RL, Baggish AL, Kovacs RJ, Maron MS. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 2: preparticipation screening for cardiovascular disease in competitive athletes: a scientific statement from the American Heart Association. *J Am Coll Cardiol* 2015;66:2356-61.
28. Gandjbakhch E, Redheuil A, Pousset F, Charron P, Frank R. Clinical diagnosis, imaging, and genetics of arrhythmogenic right ventricular cardiomyopathy/dysplasia: JACC State-of-the-Art Review. *J Am Coll Cardiol* 2018;72:784-804.
29. Dhutia H, Malhotra A, Yeo TJ, et al. Inter-rater reliability and downstream financial implications of electrocardiography screening in young athletes. *Circ Cardiovasc Qual Outcomes* 2017;10:1-9.
30. Nunes de Alencar Neto J, Baranchuk A, Bayés-Genís A, Bayés de Luna A. Arrhythmogenic right ventricular dysplasia/cardiomyopathy: an electrocardiogram-based review. *Europace* 2018;20:f3-12.

Supplementary Material

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