Cardiac Arrest and Sudden Death in Competitive Athletes with Arrhythmogenic Right Ventricular Dysplasia

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FURLANELLO F., ET AL.: Cardiac Arrest and Sudden Death in Competitive Athletes with Arrhythmogenic Right Ventricular Dysplasia. Arrhythmogenic right ventricular dysplasia (ARVD) is a predisposing factor for sport-related cardiac arrest (CA), sudden cardiac death (SD), and life-threatening ventricular tachyarrhythmias (VT). The aim of this study was the assessment of athletes with ARVD, particularly the CA survivors. From 1974 to January 1996, 1642 competitive athletes (aver. 25.5 yr.), 136 of whom were top level athletes (TLA), were studied for important arrhythmic manifestations. All athletes underwent an individualised study protocol including a series of non invasive and invasive diagnostic techniques. One hundred and one athletes (90 males, 11 females, aver. 25.9 yr.) were diagnosed as being affected by ARVD on the basis of the WHO/ISFC criteria. The same percentage (about 6%) of ARVD is present in both the general arrhythmic athletes population and in the subgroup of TLA. Prevalence of ARVD among athletes with CA or SD is high (respectively 23% and 25%), confirming the observation that ARVD is one of the major causes of SD in Italian athletes. All CA were athletic activity related, indicating the potentiality of exercise as a cause of electrical destabilisation in subjects with ARVD. In athletes with documented ARVD intense sport activity has to be proscribed. In athletes at risk of CA or SD an aggressive treatment, ICD implantation and RF catheter ablation must be taken into consideration.

ARVD, athletes, arrhythmias, cardiac arrest, sudden death

Introduction

Arrhythmogenic right ventricular dysplasia (ARVD) is a predisposing factor for sport-related sudden cardiac death (SD).¹⁻³ In fact this type of cardiopathy has been described in an increasing number of young athletes who experienced cardiac arrest (CA) or SD or in those with life-threatening ventricular tachyarrhythmias (VT). These arrhythmic manifestations can be induced by exercise. Over the past 23 years, we had the opportunity of investigating 101 consecutive competitive athletes with overt diagnosis of ARVD according to the criteria of the ARVD WHO/ISFC Task Force.⁴⁻⁵ Patients were selected from a larger population of young athletes studied for arrhythmic manifestations, including a subgroup of top level athletes (TLA).⁶⁻⁸ The aim of this paper is to study a subgroup of athletes with ARVD, who had an exercise related, during game or practice, SD and CA, comparing the prevalence of some epidemiological data with our arrhythmic athletic population and proposing a specific clinical surveillance for both the CA survivors and the other athletes after overt diagnosis of ARVD.

Methods

From 1974 to the 31st of January 1996, 1642 competitive athletes (1353 males and 289 females, of mean age 22.5 yr.) performing different types of sports,⁶⁻⁸ were referred to us from various Sport Centres, Universities and Hospitals, because of important arrhythmic manifestations (Table 1). Within these general population of competitive athletes, 136 (113 male and 23 females, mean age 23.5 yr., from 1985 to January 1996) were TLA, all competing at an international level including

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national, European, World, and Olympic champions, in 32 different competitive sports. Until referral, all athletes were considered eligible to take part in competitive sport activities according to the Italian Legislation. Each athlete underwent an individualised study protocol including a series of non invasive and invasive diagnostic techniques until a clear diagnosis of the "arrhythmic" athlete was obtained. Of the 1642 athletes, 16 (1.0%) all males, mean age 29.2 yr., had sport activity related SD. Twenty-six (1.6%), 23 males and 3 females, mean age 26.8 yr., were sent to us after being resuscitated (with CPR) for arrhythmic CA during competition or practice (Table I).

Results

Athlete Population with ARVD

Among these 1642 competitive athletes studied for arrhythmic manifestation 101 (6.1%), 90 males, 11 females, mean age 25.9 yr., were diagnosed as being affected with ARVD on the basis of the WHO/ISFC standardised diagnostic criteria consisting in major and minor clinical and structural data: two major criteria, or one major plus two minor criteria, or four minor criteria. All athletes were studied for the presence of some type of arrhythmia originating from the right ventricle (RV). The average follow up period at June 30 1997 was 98 months (min. 16 - max. 222 months). The study protocol included family and personal history, cardiac X-ray size, 12-lead ECG, 24-h ECG Holter monitoring in the basal state and during sports activity, stress test, 2-D echocardiography, electrophysiologic testing, including ventricular programmable stimulation with an aggressive protocol, contrast RV and LV cardioangiography and coronary angiography. When available, signal-averaged ECG, MRI and MIBI SPET/MBG SPET tomography studies were performed in each subject. In some cases, endocardial biopsy was performed. A complete histopathological examination was performed in 4 athletes with SD. In agreement with the COCIS 1995 exercise guidelines, all these athletes with ARVD were considered non eligible to take part in any type of competitive sport activity. The cardio-arrhythmologic surveillance of the athletes with diagnosed ARVD included a rigorous prevention of life-threatening arrhythmias and SD by means of an individualised pharmacological and/or non-pharmacological antiarrhythmic (AA) treatment. Since the beginning of our study in 1974, many new different therapeutic approaches, pharmacological and interventional, became available to treat some of these patients such as new generation ICD, implanted also in young ARVD athletes at risk of SD for intractable VT/VF. Radiofrequency catheter ablation (RFCA) of refractory ventricular tachycardia in ARVD athletes has been performed also with new technologies.

Top Level Athletes with ARVD

Nine/136 (6.6%) of TLA, all males, mean age 21.7 yr., were diagnosed ARVD after having undergone an extensive cardio-arrhythmologic protocol. Of these TLA, 2 were rugby players, 2 swimmers, 1 soccer player (European champion), 1 trotter driver (European champion), 1 basketball player, 1 martial art athlete and 1 field hockey

<table>
<thead>
<tr>
<th>Patients</th>
<th>n</th>
<th>m</th>
<th>f</th>
<th>Average age (yr.)</th>
<th>Average follow up (months)</th>
<th>n with ARVD (%)</th>
<th>n with SD (%)</th>
<th>n with CA (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmic competitive athletes</td>
<td>1642</td>
<td>1353</td>
<td>289</td>
<td>22.5</td>
<td>99</td>
<td>101 (6.1)</td>
<td>16 (1.0)</td>
<td>26 (1.6)</td>
</tr>
<tr>
<td>Top level athletes</td>
<td>136</td>
<td>113</td>
<td>23</td>
<td>23.5</td>
<td>79</td>
<td>9 (6.6)</td>
<td>1 (0.7)</td>
<td>4 (0.7)</td>
</tr>
<tr>
<td>ARVD</td>
<td>101</td>
<td>90</td>
<td>11</td>
<td>25.9</td>
<td>98</td>
<td>---</td>
<td>4 (4)</td>
<td>6 (5.9)</td>
</tr>
<tr>
<td>SD</td>
<td>16</td>
<td>16</td>
<td>0</td>
<td>29.2</td>
<td>---</td>
<td>4 (25)</td>
<td>---</td>
<td>4 (25)</td>
</tr>
<tr>
<td>CA</td>
<td>26</td>
<td>23</td>
<td>3</td>
<td>26.8</td>
<td>77</td>
<td>6 (23)</td>
<td>4 (15.4)</td>
<td>---</td>
</tr>
</tbody>
</table>

ARVD subjects with CA are described in Table II. Follow up is reported in months at 30 June 1997.
ARVD IN ATHLETES

Table II.
Clinical Data on 6 Competitive Athletes with ARVD and CA

<table>
<thead>
<tr>
<th>Pt No</th>
<th>Age</th>
<th>Sex</th>
<th>Sport</th>
<th>Premonitory symptoms</th>
<th>Circumstances of CA</th>
<th>Arrhythmias</th>
<th>ARVD</th>
<th>Treatment</th>
<th>Outcome event</th>
<th>Follow up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>27</td>
<td>M</td>
<td>Soccer</td>
<td>None</td>
<td>During</td>
<td>VT/VF</td>
<td>Diffuse form</td>
<td>Amiodarone</td>
<td>No</td>
<td>Alive (144)</td>
</tr>
<tr>
<td>2</td>
<td>30</td>
<td>M</td>
<td>Cross country skiing</td>
<td>Palpitations</td>
<td>Before</td>
<td>VT/VF</td>
<td>Diffuse form + LV involvement</td>
<td>Sotalol</td>
<td>Hyperthyroidism to amiodarone</td>
<td>Alive (100)</td>
</tr>
<tr>
<td>3</td>
<td>33</td>
<td>M</td>
<td>Soccer</td>
<td>Palpitations</td>
<td>During</td>
<td>VT/VF</td>
<td>Localised form</td>
<td>Amiodarone</td>
<td>Proarrhythmic to propafenone</td>
<td>Alive (115)</td>
</tr>
<tr>
<td>4</td>
<td>32</td>
<td>M</td>
<td>Soccer</td>
<td>None</td>
<td>During</td>
<td>VT/VF</td>
<td>Diffuse form + LV involvement</td>
<td>Sotalol</td>
<td>Hyperthyroidism to amiodarone + VT/VF CA</td>
<td>SD (75) riding motorcycle</td>
</tr>
<tr>
<td>5</td>
<td>27</td>
<td>F</td>
<td>Long distance running</td>
<td>Recurrent syncope</td>
<td>After</td>
<td>VT/VF</td>
<td>Diffuse form</td>
<td>Sotalol</td>
<td>Proarrhythmic to propafenone</td>
<td>Alive (27)</td>
</tr>
<tr>
<td>6</td>
<td>34</td>
<td>M</td>
<td>Cycling cross</td>
<td>Syncope</td>
<td>During</td>
<td>VT/VF</td>
<td>Diffuse form + LV involvement</td>
<td>Mapp ICD</td>
<td>Recurrences of fast VT</td>
<td>SD (6) asyst. postdischarge of AICD, for VT/VF</td>
</tr>
</tbody>
</table>

player. In two cases, ARVD was of the diffused form with left ventricular involvement. In three cases, the diagnosis was confirmed by endomyocardial biopsy. All were studied for documented RV tachyarrhythmias and with localised QRS abnormality depolarisation and repolarisation (right precordial leads). Two had clinical and inducible (during stress and with VPS) sustained VT (sVT), 5 complex VEBs and 2 non-sustained VT (nsVT). Three were symptomatic for severe cardiopalmus. The mean follow up period was of 79 months. All TLA (except one who dropped out of the study) are alive on AA drug treatment: sotalol in 3, nadolol in 3, amiodarone in 1, propafenone in 1.

Cardiac Arrest in ARVD Athletes

Of the 1642 athletes, 26 (1.6%) (23 males and 3 females mean age 26.8 yr.) were studied after being resuscitated (with CPR) from arrhythmic CA during competition or practice (Tables I and II), with average follow up period of 102 months at June 30 1997. Six/26 (23%) athletes, 5 males, 1 female, mean age 33.3 yr. had ARVD. Specific sport activities were soccer (3), cross country skiing (1), cycling cross (1), marathon (1, female). Prodromal exercise related symptoms were present in 4/6 athletes (66%): prolonged or severe cardiopalmus in 2, syncope in 2, not known in 2. The onset of CA was related to physical activity (organised competition or game) in all, in 1/6 before exercise (soccer game), in 4/6 during exercise (soccer game in 2, cycling cross competition in 1, cross country skiing competition in 1) and in 1/6 after exercise (marathon competition). The CA was in all cases due to RV tachyarrhythmias with VF as final arrhythmia. All these athletes were resuscitated with CPR initiated by bystanders or emergency medical technicians. The average follow up period was 77 months (min. 38 - max. 148). Three/6 athletes are alive, under antiarrhythmic drug treatment: two on sotalol, one on amiodarone. One (female) previously had major proarrhythmic effect (propafenone) during VPS (5 years after CA). One/6 dropped out. Two/6 had SD during the follow up period: both also had a deterioration of the ARVD, diffuse form and left ventricular involvement. The first patient was a competitive cyclist with ARVD and polymorphic VT refractory to drugs and mapp-guided cardiosurgical treatment, with an implanted AICD of first generation without antibradycardia.
option. He had an asystole after discharge for VF 6 months after CA. The second patient, a soccer player in therapy with sotalol, died while riding a motorcycle 75 months after the first CA. He was a “responder” to electropharmacologic testing with endocavitary PVS, performed 24 months prior to the fatal event.

Sudden Cardiac Death in ARVD Athletes

Four athletes with ARVD, all males, (4% of the 101 ARVD athletes, 25% of arrhythmic athletes SD of general population - aver. age 34.2 yr.) experienced SD (Table I). Three athletes died under antiarrhythmic treatment; two had a previous CA and they have been already described. One (previously a competitive cyclist) with diffuse form of ARVD and left ventricular involvement, died during exercise, in inadequate compliance of sotalol treatment, with numerous of nsVT and sVT episodes during follow up. One athlete (competitive soccer player), with previously documented clinical and induced polymorphic nsVT and ARVD localised form, died on field during forbidden athletic activity (while training a soccer team).

Discussion

A silent ARVD may be present in young competitive athletes, also in top level ones, that can endanger her/his sport career and in some cases lead to life-threatening arrhythmias, CA and SD. In fact, among our population of 1642 athletes studied for arrhythmic manifestations 101 (6.1%), including 6.6% of TLA, were diagnosed ARVD according to the WHO/ISFC criteria. Moreover, the prevalence in our data of ARVD in subjects succumbed to CA (23%) and/or SD (25%), confirms post-mortem observations indicating ARVD as one of the major causes of SD in Italian athletes. All CA were exercise related (before, during, after competition), confirming the documented potential of electrical destabilisation induced by physical exercise on subjects with ARVD. Also, more than 60% of these patients had important prodromal symptoms, including syncope, related to athletic activity. In all arrhythmic athletes with a suspected ARVD, a complete cardio-arrhythmologic protocol has to be performed to detect the underlying cardiopathy and evaluate the subsequent sport related arrhythmic risk of life-threatening VT, CA, and SD. In all athletes with diagnosed ARVD, and particularly in subjects survived a CA, a cardio-arrhythmologic surveillance has to be performed by means of an individualised anti-arrhythmic treatment, pharmacological and/or interventional (ICD implantation, RFCA), if necessary. To all these athletes intense sport activity has to be proscribed. Only in some minor forms of the cardiopathy and with an acceptable arrhythmic risk should a recreational physical activity be allowed.

We may have to look for a more aggressive therapeutic strategy than in the past, as in some cases of ARVD (often with left ventricle involvement) the progression of the arrhythmogenic cardiomyopathy evolves in time modifying the electrophysiological substrate and the effectiveness of the AA treatment. In fact, ARVD was previously diagnosed in four athletes succumbed to SD and two survived a CA. Other athletes with CA had VT recurrences or complained severe proarrhythmic effects due to AA drugs (Table II). Moreover, we have to be aware that several athletes may die due to insufficient compliance to the AA drug treatment and to indication of life style (e.g. if forbidden athletic activity is prosecuted). Since ARVD athletes are usually young subjects with conserved ventricular function and without presence of heart failure, it is however possible to apply a more aggressive treatment to CA survivors than to older patients.

From our experience, a favourable prognosis is seen when neither evolution of the cardiopathy nor recurrences of severe arrhythmias are detected, when there is a good compliance of the subject to the therapy under a close cardio-arrhythmologic surveillance, and if the athletic activity is really interrupted.
References

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