Medicina Sportiva (2018), vol. XIV, no 1, 3010-3016
Journal of the Romanian Sports Medicine Society

Right ventricular enlargement in a 14-year-old karate athlete – a case report

Oana Maria Corîci[1], Oana Mirea-Munteanu[2], Ionuț Donoiu[2], Octavian Istrătoaie[2], Maria Iancău[1]

[1] *Department of Physiology, University of Medicine and Pharmacy, Craiova, Romania*
[2] *Department of Cardiology, University of Medicine and Pharmacy, Craiova, Romania*

Abstract. Right ventricular (RV) dilatation is a recognized morphological adaptive change induced by exercise, but also a major echocardiographic criteria for the diagnosis of inherited cardiomyopathies such as arrhythmogenic right ventricular cardiomyopathy (ARVC). Particularly in athletes, the "grey zone", when mild phenotypes of cardiomyopathies overlap with physiological changes, poses important matter of debate.
We report a case of a 14-year-old elite karate athlete who was referred for transthoracic echocardiographic pre-participation screening and was found to have increased right ventricle dimensions. No abnormalities were identified at clinical examination. Twelve lead ECG (12-lead ECG) showed sinus rhythm (80 beats/min), QRS axis at 45 degree and duration of 120ms. Additionally, delta wave was identified in leads DI, DII, V2-V6. Transthoracic echocardiography revealed right ventricular and pulmonary artery trunk enlargement, mild tricuspid regurgitation, a mild rise in pulmonary systolic arterial pressure and moderate pulmonary regurgitation. Cardiac magnetic resonance showed enlargement of right ventricle with no identifiable abnormalities such as regional RV akinesia, dyskinesia or dyssynchronous RV. RV ejection fraction was 71%, end diastolic volume of 91 mL/m^2 and end-systolic volume of 47 ml. In conclusion, morphological cardiac consequences of long-term high intensity physical activity may fall into the grey zone, and thus becoming a challenge to distinguish the physiologic remodelling from pathological one. Therefore, our patient was advised to withdraw from the competitive sport and a close follow-up was crucial.
Key words: *athletes, RV dilation, WPW syndrome.*

Introduction

Sports activities are divided into static (isometric, resistance), dynamic (isotonic, endurance) and mixed according to the type and intensity of the exercise performed (1). Cardiac remodelling induced by sports may overlap with pathological changes seen in inherited cardiomyopathies and thus confuse the diagnosis.
Due to long term pressure overload, high-intensity static exercise was demonstrated to induce left ventricle (LV) hypertrophy and minor changes in end-diastolic and end-systolic volumes, whilst high dynamic exercise is mostly related to eccentric LV remodelling (2-3). Right ventricular (RV) enlargement is a common finding in athletes involved in high-dynamic sports whereas little evidence was noted regarding the RV changes in resistance-based athletes such as weight lifting, disc throwing, karate. Long term high-dynamic activity is associated with RV eccentric remodelling consisting in both enlargement of the cavity and increased thickness of RV wall, as a primary response to volume overload (4).
A greater degree of adaptation was documented at the right ventricle inflow (sinus) than at the outflow (infundibulum) and two-dimensional conventional echo measurements of RV function such as fractional area change (FAC) and tricuspid plane systolic excursion (TAPSE) are usually normal in athletes (4-6). A mild increase of pulmonary arterial systolic pressure (PASP) was also demonstrated in healthy endurance athletes (7). Although not included in routine pre-participation, echocardiographic evaluation of RV dimensions and morphology is extremely valuable for the exclusion of genetic cardiomyopathies such as ARVC and hypertrophic cardiomyopathy (5). Exercise has been shown to trigger the development and progression of cardiomyopathies and associated with acute changes in volume and pressure overload (8-9).
Another consequence of intensive and sustained training is electrical remodelling, associated with ECG changes that may be interpreted as normal, borderline or abnormal findings in athletes.

Wolff-Parkinson-White (WPW) is a cardiac conduction system disease characterized by the presence of an accessory pathway which directly connects the atria to the ventricle, resulting in early activation of ventricular myocardium, leading to atrial and ventricular tachy-arrhythmias with potentially life-threatening consequences. (10) Amongst athletes, a particular "WPW pattern" was described by ECG changes of WPW syndrome without the evidence of clinical symptoms. WPW pattern occurs more common in athletes than in general population, with a prevalence of approximately 1:1000. (11)
We herein describe the case of a14-year-old athlete male performing professional karate with severe right ventricular enlargement, which made challenging the differentiation between physiologic adaptation to exercise and cardiomyopathies such as arrhythmogenic right ventricular cardiomyopathy.

Case presentation
A 14-year-old karate athlete participating in competitive activities with no history of palpitation, fatigue, or documented arrhythmias, but with electrocardiographic features of Wolff-Parkinson-White Syndrome, was referred to Cardiology Research Centre of Emergency County Hospital of Craiova, for complete cardiac evaluation.
Family history was negative for any inherited cardiomyopathies or sudden cardiac death. His vital signs were normal. Physical examination demonstrated a systolic ejection murmur grade 2/6 in the left parasternal area. Routine laboratory blood tests were within limits.
A 12-lead electrocardiogram revealed sinus rhythm of 80 bpm, a short PR interval of 100ms, large duration of the QRS complex due to the presence of delta wave and inverted T waves in V1 (Figure 1, A).
Chest X-ray was normal.

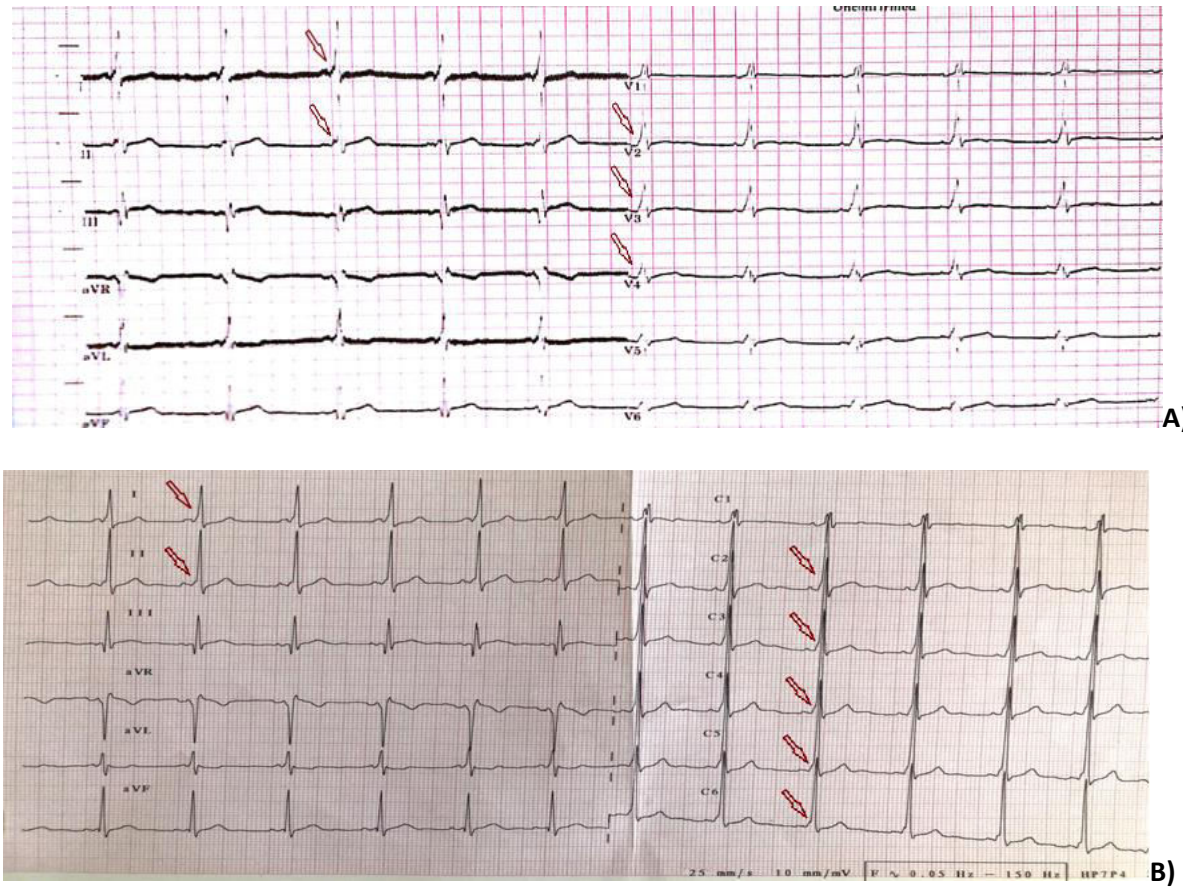


Figure 1. A) On presentation, 12 lead-ECG revealing delta waves (red arrows) B) Follow-up 12-lead ECG showing no change after 4 months of detraining.

Transthoracic echocardiography was performed using a Vivid S6 machine (GE Medical Systems-Vingmed, Horten, Norway) and showed normal left ventricular dimensions and function with an ejection fraction of 60%.

Measurement of proximal right ventricular outflow tract (RVOT) showed severe dilation (45 mm, 26 mm/m^2) which raised the suspicion of arrhythmogenic right ventricular cardiomyopathy (Figure 2, Panel A). Functional assessment using both tissue Doppler Imaging and speckle tracking echocardiography showed, however, preserved RV free wall function with a basal RV segmental velocity of 14 cm/s and RV global longitudinal strain value of -19.1. Additionally, dilated pulmonary trunk, moderate pulmonary and tricuspid regurgitation were identified. PASP was calculated 39 mmHg (Figure 3, A).

In the light of this findings, the patient was referred for cardiac magnetic resonance imaging (CMR) The examination was performed with a Siemens Magnetom Avanto 1.5 T MRI equipment (Siemens Medical Solutions USA) and confirmed right ventricle enlargement with no identifiable abnormalities such as regional RV akinesia, dyskinesia or dyssynchronous RV. RV ejection fraction was 71%, end diastolic volume of 91 mL/m^2 and end-systolic volume of 47 mL (Figure 4). Gadolinium enhancement uptake was normal in the RV myocardial regions, without myocardial fibrosis.

The patient was discharged home, with the recommendation of withdrawal from karate sports and to be reviewed after 4 months. During echo follow-up, performed at 4 months after the initial examination we could determine a decrease in RV end-diastolic dimensions (from 45 mm to 34 mm) (Figure 2, Panel B), but no significant change in tricuspid and pulmonary regurgitation grading and PASP (37 mmHg) (Figure 3, B), 12-lead ECG tracing was unchanged (Figure 1, B).

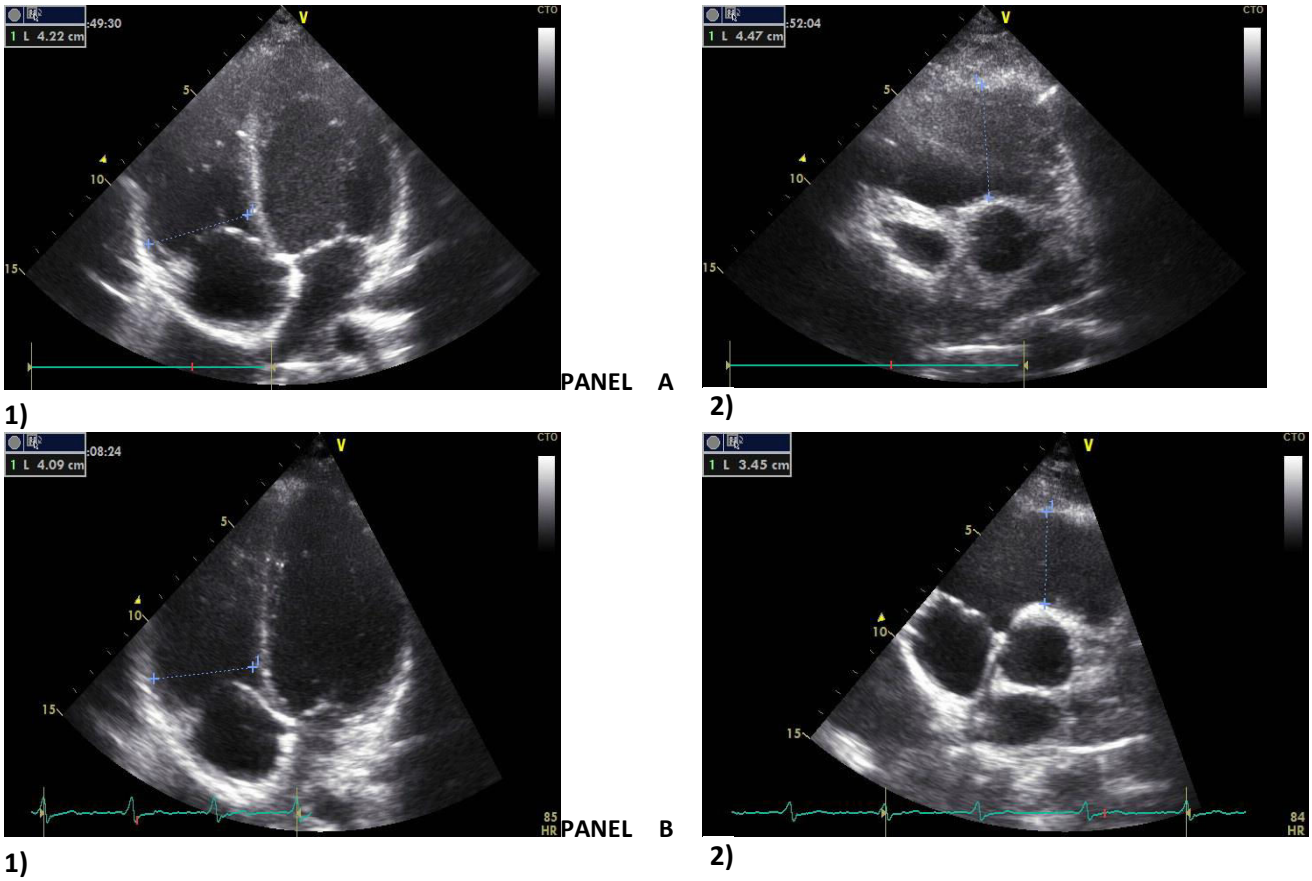
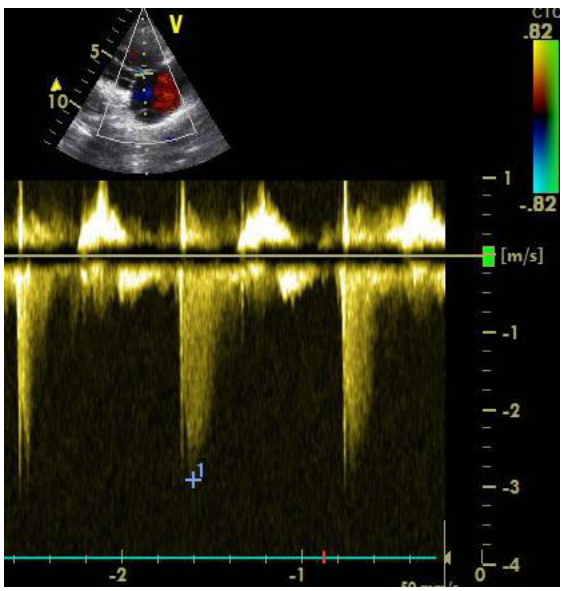
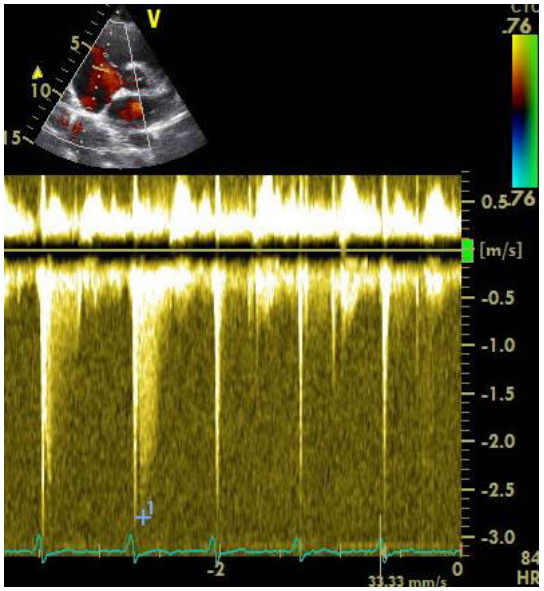


Figure 2. Transthoracic echocardiography (TTE) 1) RVOT basal diameter (mm). 2) RVOT proximal diameter (mm) - Panel A. First echocardiographic assessment, RVOT basal and proximal dimensions showing a dilated right ventricle. Panel B. Follow-up assessment showed a decrease in RV end-diastolic dimensions from 45 mm to 34 mm.

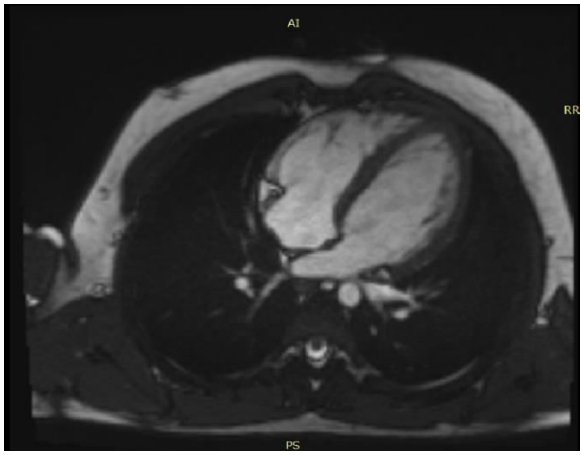


A)

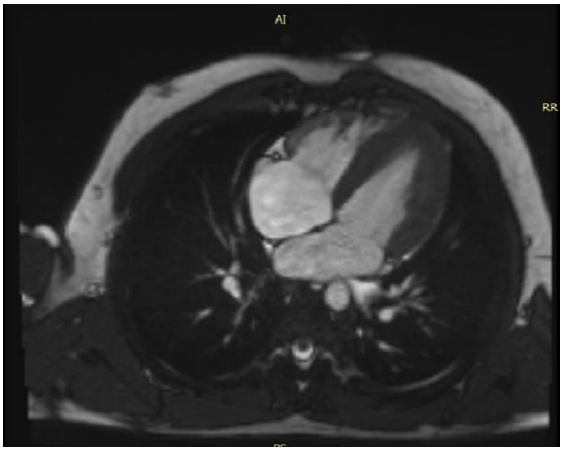


B)

Figure 3. Transthoracic echocardiography. No significant change in PASP (mmHg) between first echocardiographic and follow-up assessment.



A)



B)

Figure 4. Cardiovascular Magnetic Resonance (CMR) (4-chamber view). Cine images in end-diastole (A) and end-systole (B) showing right ventricular enlargement with an end diastolic volume of 91 mL/m^2 and no identifiable abnormalities.

Discussion

According to the type and intensity of the exercise performed, sports activities are divided into static (isometric, resistance), dynamic (isotonic, endurance) and mixed (1). Intensive exercise was associated with the development of both electrical and structural cardiac changes which, in certain situations, may overlap with inherited cardiomyopathies making thus challenging the differentiation of "grey-zone" exercise-induced structural adaptations from pathologic remodelling (13-14). The morphologic and functional changes determined by intensive exercise have been extensively studied (15-16) and include enlargement of ventricular cavities with preserved function (12) (endurance activity) and left ventricular concentric remodelling (resistance activity). Right ventricular dilatation is a common finding in athletes performing high intensity endurance exercise and in certain situations of excessive remodelling the differential diagnostic with inherited cardiomyopathies may become clinically challenging.

We presented the case of a young athlete performing karate (static) who was found to have severe dilatation of right ventricle, pulmonary artery trunk enlargement, mild tricuspid regurgitation, a mild rise in pulmonary

systolic artery pressure and moderate pulmonary regurgitation. Additionally, he presented electrocardiographic features of Wolff-Parkinson-White Syndrome.
The case posed important dilemmas as karate was classified a high-static demand training with reduced structural and functional consequences on the RV.
Apart from a characteristic of sports, RV enlargement is also considered a major echocardiographic criteria for the diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC), which is the most common differential diagnosis. Therefore, in our case the justification for further assessment of right ventricular structure was based on confirming or excluding this pathology which is a life threatening genetically inherited disease, being attributed to 4-22% of SCD (17-18). It is characterized by fibro fatty infiltration of normal cardiac myocytes (14) which leads to right ventricular structural remodelling and systolic dysfunction and a high risk of malignant ventricular arrhythmias.
The natural history of ARVC is considered to include four distinct phases. If in advanced stages the diagnosis can be more rapidly established, the early clinical phase is characterized only by ECG abnormalities and RV-originating tachyarrhythmia during adolescence, in the absence of structural anomalies. Sudden death is often the first symptom during this phase; therefore, early non-invasive diagnosis is important.
Early diagnosis of ARVC is challenging owing to the nonspecific clinical presentation (8-9). The current guidelines proposed a set of diagnostic criteria including major and minor criteria in 6 categories: 1) global or regional dysfunction and structural alterations, 2) tissue characterization of the wall, 3) repolarization abnormalities, 4) depolarization or conduction abnormalities, 5) arrhythmias, and 6) family history.
The presence of two major, one major and two minor, or four minor criteria is considered definite diagnostic. (19-20). Whereas these criteria are highly specific, they lack sensitivity for early disease (21).
In most of the cases advanced imaging techniques such as cardiovascular magnetic resonance is required for the evaluation of potential tissue abnormalities.
Our athlete was referred for CMR which confirmed the enlargement of right ventricle with no identifiable abnormalities such as regional RV akinesia, dyskinesia or dyssynchronous RV.
Hence, in this young male only one major echocardiographic criteria for ARVC was met thus definite diagnosis for this pathology was not fulfilled.
To make things more challenging, in 2012, La Gerche et al. documented a group of endurance athletes with ventricular arrhythmias RV-originating in whom intensive endurance exercise may cause impairment of the myocardial interstitial matrix mimicking an ARVC phenocopy. Genetic analysis demonstrated a low incidence (12.8%) of a specific desmosomal gene mutation in this cohort, when compared with the prevalence of desmosomal gene mutations in series describing familial ARVC probands (30-50%).
Thus, intense physical exertion may itself cause and accelerate an acquired form of cardiomyopathy (22). No evidence was found, however, regarding the involvement of high-static training in the development of the acquired form of ARVC (4, 22).
In view of this, our patient was advised to withdraw from competitive sport to establish the implication of exercise in the development of right ventricular enlargement. At the follow-up date, we noticed a decrease in right ventricular proximal diameters of 25 %, but no significant change in tricuspid and pulmonary regurgitation grading and PASP, highlighting that even static exercise is a factor in the progression of RV cardiac structural changes.
Moreover, in our patient was noted the presence of WPW pattern. Electrical abnormalities such as profound bradyarrhythmia, supraventricular and complex ventricular arrhythmias (VA) were described in athletes. (23) Nonetheless, the incidence of death appears to be low (1 in 355 athletes during an 8-year follow-up) in athletes presenting with frequent and complex ventricular tachyarrhythmias. (24).
Wolff Parkinson White syndrome may be responsible for the occurrence of ventricular tachyarrhythmias and sudden cardiac arrest in athletes. (11) The diagnosis is established by the classic triad: a short PR interval <120ms, the presence of delta wave and QRS complex duration >120ms (25). Nevertheless, the main majority of athletes with WPW have normal cardiac structure.
The presence of clinical manifestations differentiates between the “Wolff-Parkinson-White pattern”, also known as ECG evidence of WPW without symptoms from WPW syndrome itself. When symptoms are present, the patient should be referred for ablation to eliminate the pathway, whilst in the absence of symptoms, a complete clinical evaluation including transthoracic echocardiogram (in order to establish the

potential presence of Ebstein disorder or associated cardiomyopathy) and an exercise treadmill test are recommended.
The latter is of particular importance dictating future management considering that the disappearing of the accessory pathway during the exercise is associated with low risk for SCD, and sports activity is allowed. (26). In our case, during follow-up date, we established that the WPW pattern was present as the ECG tracing was unchanged and the patient was still asymptomatic. The young athlete needs further exercise testing to evaluate whether he either continue or avoid vigorous exercise.
We recommended that the young athlete should undergo lifelong clinical follow-up (every 12 months) including 12-lead ECG, echocardiography in view to periodically evaluate new-onset symptoms and progression of morphological remodelling. In view of this, in athletes an accurate diagnosis is crucial as an erroneous diagnosis carries serious risks, leading to either adverse events or disqualification from sport, with financial and psychological implications.

Conclusion

The current case described the association of WPW pattern with right ventricular enlargement in an athlete practicing karate. Our patient's investigations placed him in the "grey zone" noticing the presence of right ventricular dilation, pulmonary trunk enlargement with the presence of tricuspid regurgitation, but all the parameters were framed as borderline and was met only one major criteria for inherited cardiomyopathy. At the follow up date, after 4 months of detraining, there was a decrease in right ventricular diameters, highlighting the role of exercise in the development of cardiac structural changes. WPW pattern was unchanged. In conclusion, our case was a challenge in establishing the real boundaries between physiological cardiovascular features related to sports and pathological remodeling, in the lack of normative athletic cardiac references/values.

References

1. Mitchell JH, Haskell W, Snell P, Van Camp. SP. (2005) Task Force 8: Classification of Sports. *JACC* V; 45 (8): 1364-7.
2. Mitchell JH, Raven PB (1994). Cardiovascular adaptation to physical activity. In: Bouchard C, Shephard R, Stephen T, editors. *Physical Activity, Fitness, and Health: International Proceedings and Consensus Statement*. Champaign, IL: Human Kinetics: 286 –98.
3. Gallagher KM, Raven PB, Mitchell JH. (1999) Classification of sports and the athlete's heart. In: Williams RA, editor. *The Athlete and Heart Disease: Diagnosis, Evaluation and Management. Philadelphia*, PA: Lippincott Williams & Wilkins: 9 –21.
4. D'Andrea A., Riegler L., Golia E., et al. (2013). Range of right heart measurements in top-level athletes: the training impact. *Int J Cardiol*; 164:48-57.
5. Oxborough D., Sharma S., Shave R., et al. (2012). The right ventricle of the endurance athlete: the relationship between morphology and deformation. *J Am Soc Echocardiogr*; 25: 263-71.
6. Zaidi A., Ghani S., Sharma et al. (2013) Physiological right ventricular adaptation in elite athletes of African and Afro-Caribbean origin. *Circulation*; 127: 1783-92.
7. D'Andrea A, Naeije R, D'Alto M, Argiento P et al. (2011). Range in pulmonary artery systolic pressure among highly trained athletes. *Chest*. 139(4): 788-794.
8. Basso C, Corrado D, Marcus F, et al. (2009). Arrhythmogenic right ventricular cardiomyopathy. *Lancet*; 373: 1289-300.
9. Basso C., Bauce B., Corrado D., and Thiene G (2012). Pathophysiology of arrhythmogenic cardiomyopathy. *Nat Rev Cardiol*; 9(4): 223-33.
10. Wolff L., Parkinson J., and White P. (1930). Bundle-branch block with short PR interval in healthy young people to paroxysmal tachycardia. *Am Heart J V*: 685-704.
11. Pelliccia A., Culasso F., Di Paolo F.M. et al. (2007). Prevalence of abnormal electrocardiograms in a large, unselected population undergoing pre-participation cardiovascular screening. *Eur Heart J*; 28: 2006-10.
12. Bauce B., Frigo G., Benini G., et al. (2010) Differences and similarities between arrhythmogenic right ventricular cardiomyopathy and athlete's heart adaptations. *Br J Sports Med*; 44(2):148-54.
13. Kim JH, Baggish AL. (2016) Differentiating exercise-induced cardiac adaptations from cardiac pathology: the "grey zone" of clinical uncertainty. Can J Cardiol; 32(4): 429-37.
14. Jeffrey S. Hedley, Ibhar Al Mheid, Zoubin Alikhani, Maria A. Pernetz, Jonathan H. Kim. (2017). Arrhythmogenic Right Ventricular Cardiomyopathy in an Endurance Athlete Presenting with Ventricular Tachycardia and Normal Right Ventricular Function. *Tex Heart Inst J*; 44(4):290-3.

15. Pavlik G et al. (2013). Acta Physiol Hung. The athlete's heart. Part II: influencing factors on the athlete's heart: types of sports and age (review). *Acta Physiol Hung Mar*; 100(1):1-27.
16. Shavit R Glikson M, Constantini N. (2016). Athlete's heart - the more the merrier? *Harefuah*; 155(9):531-536.
17. Corrado, D., Basso, C., Schiavon, M. and Thiene, G. (1998). Screening for hypertrophic cardiomyopathy in young athletes. *New Engl J Med*; 339(6): 364–9.
18. Corrado D., Basso C, Pavel A., et al (2006). Trends in sudden cardiovascular death in young competitive athletes after implementation of a preparticipation screening program. *JAMA*; 296 (13): 1593-601.
19. McKenna WJ, Thiene G, Nava A, Fontaliran F, Blomstrom- Lundqvist C, Fontaine G, et al. (1994). Task Force of the Working Group Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology. Diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy. *Br Heart J*; 71(3): 215-218.
20. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, et al. (2010). Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. *Circulation*; 121(13):1533-1541.
21. Sen-Chowdhry S, Lowe M, Sporton S, McKenna WJ. (2004) Arrhythmogenic right ventricular cardiomyopathy: clinical presentation, diagnosis, and management. *Am J Med;* 117:685–95.
22. La Gerche A., Robberecht C., Kuiperi C. et al. (2010). Lower than expected desmosomal gene mutation prevalence in endurance athletes with complex ventricular arrhythmias of right ventricular origin. *Heart*; 96 (16): 1268-74.
23. Whyte GP, Stephen N., Senior R., Peters N. et al. (2008). Differentiation of RVOT-VT and ARVC in an Elite Athlete. *Journal of American College of Sports Medicine*.
24. Biffi A, Pelliccia A, Verdile L, et al. (2002). Long-term clinical significance of frequent and complex ventricular tachyarrhythmias in trained athletes. *J Am Coll Cardiol*; 40:446–52.
25. Surawicz B., Childers R, Deal BJ, et al. (2009). AHA/ACCF/HRS recommendations for the standardization and interpretation of the electrocardiogram: part III: intraventricular conduction disturbances: a scientific statement from the American Heart Association Electrocardiography and Arrythmias Comittee, Council on Clinical Cardiology; the American College of Cardiology Foundation; and the heart Rhythm Society: endorsed by the International Society for Computerized Electrocardiology. *Circulation*; 119(10): e235-40.
26. Cohen MI, Triedman JK, Cannon BC, et al. (2012). PACES/HRS expert consensus statement on the management of the asymptomatic young patient with a Wolff-Parkinson-White (WPW, ventricular preexcitation) electrocardiographic pattern. *Heart Rhythm*; 9: 1006-24.

Corresponding author
Oana-Mirea Munteanu
Department of Cardiology, University of Medicine and Pharmacy Craiova, Romania
Phone: +40723287000
E-mail: *oana.mirea83@gmail.com*

Received: February, 2018
Accepted: April, 2018