

ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA – CAUSE OF DEATH OF YOUNG ATHLETE (STUDY OF CASES)

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Abstract

In communication is presented a case of the death of an adolescent soccer player due to arrhythmogenic right ventricular dysplasia. Five years before the death were found by athlete during preventive medicine examination some changes on ECG (negative T-waves over the right precordium and disorders of rhythm) and he was sent to pediatric cardiologist. The cardiologist has confirmed negative T-waves and has recommended following. But athlete didn't come for control any more and last four years before death he didn't come for preventive medical check-up at all.

We suppose, that resting and loading ECG examination during the preventive check-up of athlete by the physician of physical training has its justification. For the physicians of physical training we remind the meaning of careful anamnesis, clinical examination and resting and loading ECG in disclosure of this disease. From the knowledge of ARVD and of our case follows a recommendation to the athletes, their parents and sport pedagogues, that they did not underrate the risk of myocarditis and cardiomyopathy, that they pass preventive medical check-up and that they respect the recommendation of physicians. We recommend to the Universities to deal with this problems during lessons of the sport pedagogues and physicians.

Key words: heart disease, sudden death, sport

Case history

Clinical course:

In the late 2007 during the indoor soccer match in South Moravia has suddenly collapsed 16 years old soccer player. He was immediately resuscitated by laymen. Called Emergency Medical Service carried out advanced cardiopulmonary resuscitation. The boy was transported to the clinic of children anaesthesiology and resuscitation. After 1 hour and 20 minutes of resuscitation came to death. Health dissection was recommended.

Results of health dissection:

By external examination on pathology was described a cyanosis of the face, oedematous fluid in nasal and oral cavity, reasonable gynaecomastia and as well obesity. Dissection finding had following statements: considerable congestion and expressive pulmonary oedema, cardiac hypertrophy with right-sided dilatation (Fig.1), abnormal impair of musculature of right ventricle and its substitution through adipoid tissue, what is considerable already grossly (Fig.2). Microscopical picture has proved oedema of brain, acute venostasis, haemorrhagic intra-alveolar pulmonary oedema. On the section of right ventricle (Fig. 3) was found a diffusion multiplication of mature and immature ligament with creation of focus,

multiplication of adipoid tissue perivascularly also among cardiomyocytes, compensatory hypertrophy of cardiomyocytes. It was a diffusion replacement of right ventricle myocardium by ligament adipoid tissue. In the tissue from left ventricle was described hypertrophy of cardiomyocytes and congestion of vessels. **In conclusion** of health dissection was named as a direct cause of death arrhythmogenic right ventricle cardiomyopathy (dysplasia).

Results of examination during preventive medicine exams by sports physician:

During the years 2000-2004 was athlete submitted to medicine examen of physical training (Tab. 1).

In personal and family anamnesis hasn't occurred any serious diseases. The boy was without subjective troubles.

Objectively was found gradually raising body mass index by examinations, adipoid component – slowly developing mild obesity, approximately average vital capacity of lungs, mild ingravescent working capacity according to re-count of weight (W_{170} /kg). It was described mild disorder of body hold and mild muscular imbalance. Already since the first examination in ECG were described nonspecific and quite usual changes: negative T waves in lead III and in the leads from right precordium. In January 2003 were found

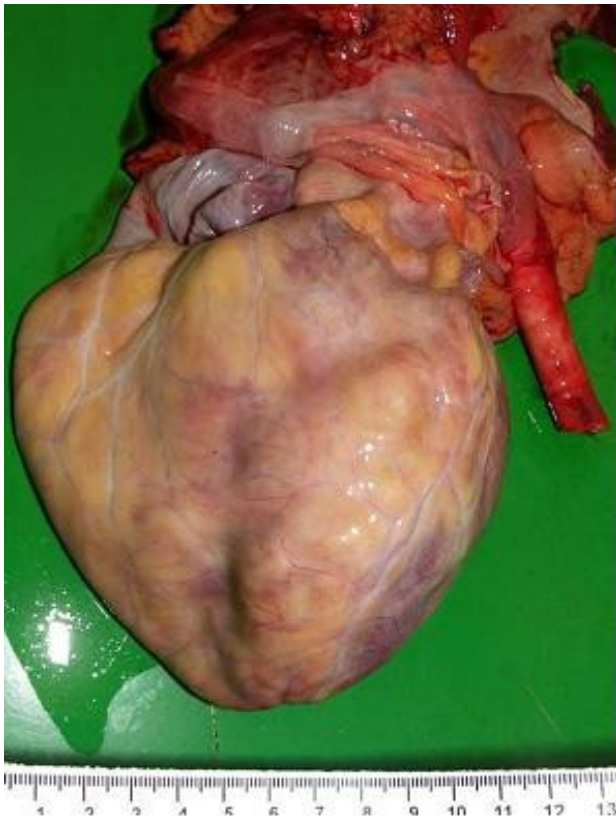


Fig.1. The cardiac finding of right ventricle dilatation (Photographer: Jiří Horák). Look at anterior side

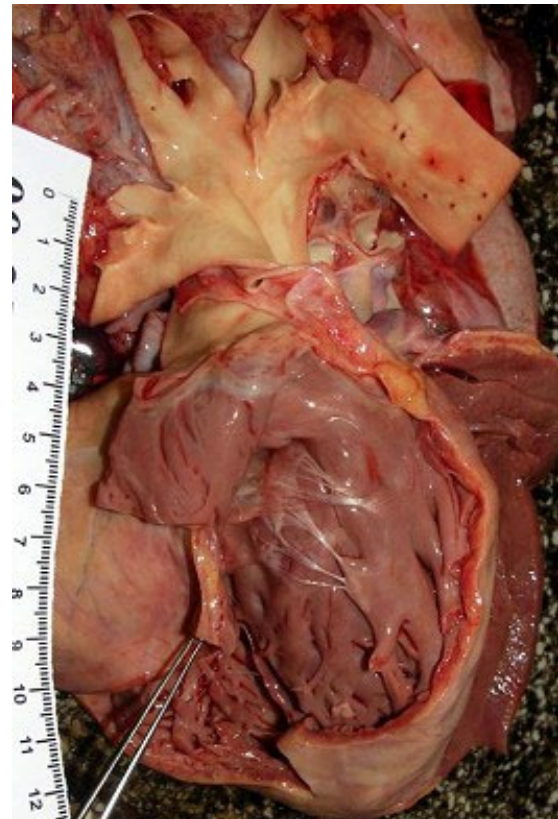


Fig.2. The cardiac finding of depletion and apoptosis of right ventricular wall (Photographer: Jiří Horák)

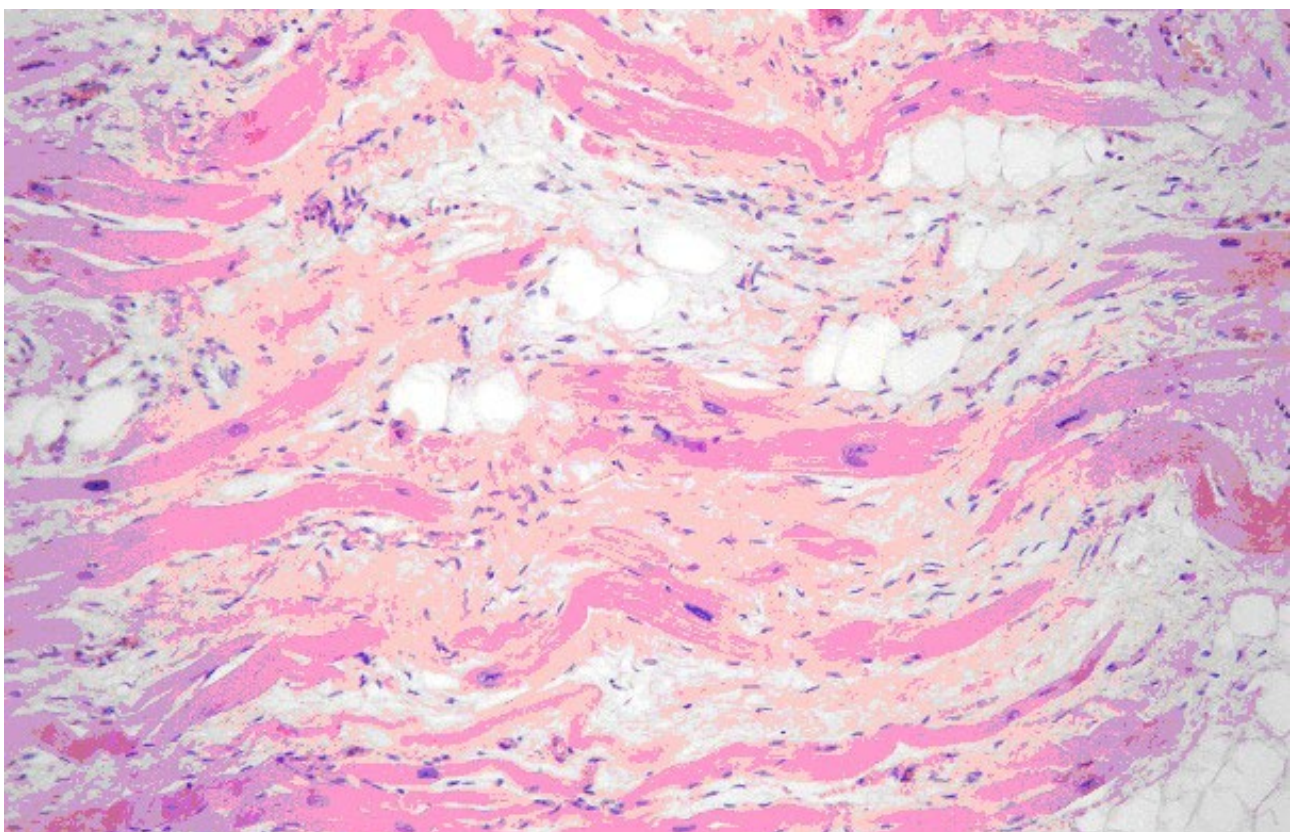


Fig.3. Microscope finding in myocardium of right ventricle (Photographer: Jiří Horák)

Table 1. *The results of preventive sports medicine exams*

Time / Age (yrs)	Dec 2000 / 9.5	Dec 2001 / 10.5	Jan 2003 / 11.5	Feb 2004 / 12.5
Anamnesis	None serious diseases, injuries and operations, without troubles			
Sport history	Football 5x a week 1.5 hour			
Weight Hight	41 kg 141 cm	44 kg 146 cm	51 kg 152 cm	52 kg 156 cm
BMI Fat	20.6 25%	20.6 24%	22.1 25%	21.4 28%
FEVC	2.5 l 104%N	2.6 l 98%N	2.8 l 95%N	3.1 l 98%N
PWC170/kg	2.65 W 120%N	2.13 W 95%N	2.20 W 94%N	2.00 W 82%N
Status praesens	incip. obesity light FDP, CP physiol.	FDP, CP physiol.	obesity, FMD	FDP, obesity, FMD, CP physiol.
Rest and exercise ECG	neg. T in III and V1-2	neg. T in V1SHA	marked arrhythmia	neg. T in III and V1-4
Recommendation	on obesity	on obesity	on obesity ad cardiologist	on obesity

Legend: %N - percentage of reference value, BMI - body mass index, FEVC - forced expiratory vital capacity of lungs, PWC170/kg - power work capacity on heart rate 170/min per kilogram of body mass, ECG - electrocardiogram, FDP - functional disorder of posture, CP physiol - normal clinical status of cardiopulmonary system, L - lumbal, MD -functional muscle imbalance, neg. - negative, T - T wave, III - III. limb lead, V1 - 1 st chest lead, SHA - sinus heart arrhythmia

already suspicious disorders of heart rhythm, which were no more considered as only physiologic sinusoidal arrhythmia and the boy was sent to the children cardiologist for further examination (see next). In 2004 he came to preventive medicine examen for the last time, i.e. almost 4 years before death.

The result of examination by children cardiologist in January 2003:

Regarding anamnesis the boy was without troubles, he didn't feel any irregularity of heart, he was equal with his team mates. His father had a high blood pressure. His grandfather has died because of heart trouble, but it wasn't sudden death. Objectively was found normal filling with blood, pulse in tan, non-enlarged liver. Listening finding on heart: no vortex, regular action, heart sounds normal demarcated, without murmur. Electrocardiography at rest lying: heart action 60/min, sinusoidal rhythm, axis QRS plus 30 degrees, PQ 0.14 s, QT 0.40 s, negative T wave in leads V1-3, T positive from V4, in V6 has a complex QRS shape qRs, without hypertrophy symptoms. Echocardiography at rest lying: concavity of atrial diaphragm in the middle part towards the right atrium, but without any symptoms of shunt, normal motion of septum, there is no enlargement of heart sections, normal anatomy and deposit of heart, normal flows, normal valves, ejection fraction of left ventricle 67%, heart rate 51-63/min, sinusoidal dysrhythmia, inclination to bradycardia. **Conclusion and recommendation:** demarcated disorder of heart rhythm by the young recreative athlete. It is necessary to control pulse (on hand), when decreasing under 50/min or by irregularity - examen at cardiologist immediately.

Discussion

Physician of physical training has sent correctly athlete to cardiologist when remarking suspicious disorders on ECG screening. The physician didn't find any further symptoms of pathology and recommended selfcontrol and in case of disorders of rhythm examen at cardiologist immediately. In that time fulfilled the changes in ECG screening only one small criterion for determination of ARVD diagnosis (negative T waves in right precordium). But the athlete didn't come to any further examen to cardiologist. Neither by the last examen at sports physician, almost 4 years before the death, weren't found clear symptoms of pathology. We suppose, that our athlete hasn't come to the examen to cardiologist and consequentially he also stopped to come once a year to preventive examen to sports physician in connection with his running puberty (age 13-16 years). Very difficult diagnostics in early stages of disease also demands repeated examination. Especially in cases of subclinical phase of ARVD disease, there is very important using of diagnostic apparatus. According to Marek (2003) [1] has its place in ARVD diagnostics also ergometry (loading ECG test), because by the young athletes arise the ventricular tachycardia during exertion.

As the physician of physical training can do in his surgery resting and also ECG stress test, we mention the most often described changes in ECG screening: *widening of QRS complex over the right precordium* [2, 3]. Composite authors of John Hopkins Hospital [4] recommends to count ratio of intervals of QRS in V1-V3 to QRS in V4-V6. 97% of cases is ARVD greater than 1.2. But sensitiveness of this QRS diagnostic criterion

wasn't determined. *Ventricular extrasystoles and tachycardia* shaped as a block of right Tawar crus [2, 5, 3, 6]. *Presence of Epsilon wave* [3, 5, 7]. For better screening of the Epsilon wave John Hopkins Hospital (2008) recommends certain shifting of electrodes: Electrode **L** to shift over the processus xifoideus, electrode **R** over the manubrium sterni and electrode **F** into the places for V4-5. *Abnormal repolarization in the right precordium leads*, i.e. negative T waves in leads V1-V3 [3-5]. Development of changes in ECG screening should have been considered from the long-term view – during several years: In resting ECG lying it is gradual extension of negative T waves from the lead V1-2 up to V3-4 and gradual change of electric axis of QRS complex in leads of limbs in the sense from the left to the right (e.g. from plus 30 dg. Up to plus 60 or plus 90 dg.) etc.

Next examinations are already in competence of cardiologist or of further specialists: Echocardiography, magnetic resonance imaging (MRI) and computer tomography (CT) could disclose *extension (dilatation) of antrum of right ventricle* [3, 5, 8]. Especially MRI is considered as golden standard of diagnostics of this disease. But at the beginning of this disease there are no changes which could be simply demonstrable [3, 8].

Conclusions

It has been described the case of death of young athlete, who hadn't any serious health troubles up to that time. The main cause was hereditary disease of arrhythmogenic right ventricle dysplasia. There weren't found any further factors, which could share in starting of disease, e.g. myocarditis. In this case physician of physical training has found suspicious changes on ECG during preventive examination nearly 5 years before death and correctly sent an athlete to the cardiologist. But in that time cardiologist couldn't determine any ARVD diagnosis, the symptoms didn't fulfil criterions. The athlete didn't already come to any further examinations. We can theoretically to allow only that, if the athlete would visit systematic examinations by cardiologist, development of disease would probably bring next and more expressive symptoms, which would enable determination of diagnosis. But even in this case the prognosis would be very unsure.

We suppose, that both resting and stress ECG test during preventive examination of the athlete at physician of physical training has its foundation, there could be quite other situation in Europe than in the USA. We propose to create an European national registry of cases regarding sudden cardiac death of athletes, which would enable better study of this problems. Is there a possibility e.g. of widening of already existing Netherlands national registry?

Recommendations for physicians of physical training and for sports physicians:

- Carefully examine the athlete during examination – anamnesis, clinical examination, resting and loading ECG.
- Send him to cardiologist in case that the symptom will occur, to control, if the athlete keeps his recommendation.

Recommendations for the young athletes, their parents and coaches:

- Don't underestimate risk of myocarditis and cardiomyopathy.
- Go through regular preventive medicine examinations.
- Keep recommendations of physicians.

Recommendations for teachers of physicians and sports schoolmasters:

- Classify this problems among the lessons.

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Acknowledgment

We thank our colleagues Jana Hlaváčová and Vojtěch Weinberger and photographer Jiří Horák for a kind collaboration.

Received: April 02, 2010

Accepted: November 22, 2010

Published: November 30, 2010

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A – Study Design

B – Data Collection

C – Statistical Analysis

D – Data Interpretation

E – Manuscript Preparation

F – Literature Search

G – Funds Collection