Clinical Profile and Prognosis of Patients with Right Ventricular Dilated Cardiomyopathy: Results of a Prospective Study

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Abstract

The aim of our study was to investigate the clinical prevalence of dilated cardiomyopathy (DCM) with predominantly failure of the right-side heart (right ventricular DCM, RV-DCM), and features of the clinical course and prognosis of the disease compared to DCM with biventricular heart failure (BV-HF).

The study design suggests a prospective observation of 300 patients with idiopathic DCM between 2000 and 2012. Herewith, we followed the criteria of the WHO/ISFC task force (1995) on the definition and classification of cardiomyopathies. All patients underwent a comprehensive examination. Two groups were formed for further comparative analysis. Group 1 included 22 patients (mean age 42.9±14.3 years, male/female 5/17) with RV-DCM. Group 2 included 38 patients (mean age 43.6±13.8, male/female 29/9) with DCM and BV-HF. The groups were matched for age, NYHA class II-III, and disease duration. According to our aim, we studied 5-year survival prognosis and analyzed the incidence and causes of deaths, as well as the occurrence of nonfatal complications of the disease. Medical therapy for DCM patients was performed according to the CHF therapy guidelines (ACC/AHA 2001, 2005). The results of our investigations during many years of research have shown that the clinical incidence of RV-DCM was 7.3% among all forms of DCM. The study of life prognosis in patients with 2 forms of DCM showed that 5-year mortality of patients was about 50%. Fatal pulmonary embolism was a leading cause (50%) in RV-DCM patients. (*Int J Biomed.* 2015;5(4):207-213.)

Keywords: *right ventricular dilated cardiomyopathy, biventricular heart failure, prognosis; pulmonary embolism.*

Abbreviations

RV-DCM, right ventricular dilated cardiomyopathy; **CHF**, chronic heart failure; **BV-HF**, biventricular heart failure; **LVM**, left ventricular mass; **LVEDD**, LV end-diastolic dimension; **LVESD**, LV end-systolic dimension; **LVEDV**, LV end-diastolic volume; **LVESV**, LV end-systolic volume; **LVSV**, LV stroke volume; **LVFS**, LV fractional shortening; **LA** left atrium; **RA**, right atrium; **IVST**, interventricular septal thickness; **LVPWT**, LV posterior wall thickness; **EF**, ejection fraction; **ARVD**, arrhythmogenic RV dysplasia; **AF**, atrial fibrillation; **PEmb**, pulmonary embolism; **PE**, peak early filling velocity; **PA**, peak atrial filling velocity; **LBBB**, left bundle branch block; **RBBB**, right bundle branch block.

Introduction

The idiopathic nature of right ventricle failure was first described by A. Bahler in 1976 [1]. Describing two cases of RV dilatation and paradoxical movement of the interventricular septum in the absence of any disease or heart disease, the authors suggested that there are certain initial symptoms of

cardiomyopathy. Afterwards, the report of WHO/ISFC task force defined cardiomyopathies as diseases of the myocardium associated with cardiac dysfunction and classified the forms of diseases, including DCM with its varieties [2]. The international recommendations to date have provided clear criteria for the diagnosis of alcoholic, peripartum cardiomyopathy and family forms of cardiomyopathy [3-5]. There are a lot of controversial issues related to both the diagnosis and treatment of DCM with predominantly failure of the right ventricle (not to be confused with ARVD). Isolated idiopathic RV-DCM is an equally rare condition like the

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familial RV-DCM. Besides the few reports which had been mentioned casually, there are only a few documented cases in the literature. [6-9]. The severity of the problem is heightened by a significant limitation of publications related to the clinical course and prognosis precisely of RV-DCM.

Based on the above, the aim of our study was to investigate the clinical prevalence of DCM with predominantly failure of the right-side heart (RV-DCM), and features of the clinical course and prognosis of the disease compared to DCM with BV-HF.

Materials and Methods

The study design suggests a prospective observation of 300 patients with idiopathic DCM between 2000 and 2012 in the Department of Heart Failure and Noncoronary Myocardial Pathology at RSCC. Herewith, we followed the criteria of the WHO/ISFC Task Force (1995) on the Definition and Classification of Cardiomyopathies [2]. RV-DCM was diagnosed according to the above criteria and the additional exclusion of other conditions characterized by dilatation and right-sided heart failure (primarily ARVD, idiopathic and secondary pulmonary hypertension), as well as clinical, electrocardiographic, and echocardiographic features that were systematized later and proposed as criteria for diagnosis [3]. In addition, to assess the clinical status, the patients underwent 6-minute walk test (6MWT), a standard electrocardiogram («Cardio Lab», Ukraine), transthoracic echocardiography (Siemens Sonoline Verso Pro) at rest by common methods to determine the volumetric and linear heart parameters with calculation of LV myocardial contractility by Teicholz method. We used R. Levine's, T. Gibson's at al. formula: $RVEF\% = (RVEDV - RVESV) / RWEDV \cdot 100\%$ for the calculation of RVEF [10,11]. Biochemical and immune – enzyme analyses were also carried out.

In the course of examination of patients, two groups were formed for further comparative analysis. Group 1 included 22 patients (mean age 42.9 ± 14.3 years, male/female 5/17) with RV-DCM. Group 2 included 38 patients (mean age 43.6 ± 13.8 , male/female 29/9) with DCM and BV-HF. The groups were matched for age, NYHA class II-III, and disease duration. According to our aim, we studied 5-year survival prognosis and analyzed the incidence and causes of deaths, as well as the occurrence of nonfatal complications of the disease.

Medical therapy for DCM patients was performed according to the ACC/AHA 2001, 2005 Guidelines.

Statistical analysis was performed using the statistical software «Biostatics» (v.4.03). Baseline characteristics were summarized as frequencies and percentages for categorical variables and as mean $\pm$ SEM for continuous variables. Student's unpaired and paired t-tests were used to compare two groups for data with normal distribution. Group comparisons with respect to categorical variables are performed using chi-square tests or, alternatively, Yates' χ^2 when expected cell counts were less than 5. A probability value of $P < 0.05$ was considered statistically significant.

The study was conducted in accordance with ethical principles of the Declaration of Helsinki and approved by

the Republican Specialized Center of Cardiology Ethics Committee. Written informed consent was obtained from each patient.

Results

During the study period, we had the opportunity to assess the clinical and hemodynamic parameters of the patients; readmissions frequency and outcomes were recorded, including an active remote observation. The initial patients' characteristics—clinical and functional, anamnestic, gender and hemodynamic—are shown in Table 1. The vast majority of our patients (77%) identified with RV-DCM were female, while almost all epidemiological studies suggest that DCM as a whole is more common in men. The patients' mean age (more than 40 years), as well as the age when the disease manifested, was comparable in both groups.

Table 1.

The initial patients' characteristics

Variable	Group 1 (n=22)	Group 2 (n=38)	P
Average age, years	42.9 ± 14.3	43.6 ± 13.8	NS
Male/female	6/16	29/9	<0.05
Average age for disease onset, years	38.2 ± 14	38.7 ± 14	NS
Diagnosis of DCM during the primary examination	2 (9%)	8 (21%)	NS

Among RV-DCM patients during the primary examination, DCM was diagnosed only in 9% of cases vs. 21% of cases among DCM patients with BV-HF ($P < 0.05$). The higher percentage among patients with BV-HF was possibly caused by obvious signs of LV failure and the detectable LV dilatation. The majority of Group 1 patients were treated and observed with other diagnoses, such as chronic hepatitis, COPD, coronary heart disease, liver cirrhosis, etc.

Clinical evaluation of the patients revealed some features that depended on the character of myocardial failure. We analyzed the symptom scores to assess the differences (Table 2) between Groups.

Table 2.

Clinical characteristics of the studied patients

Variable	Group 1 (n=22)	Group 2 (n=38)	P
Dyspnea	1.35 ± 0.49	1.42 ± 0.51	0.614
Patient's position in the bed	0.89 ± 0.16	1.58 ± 0.51	0.000
Jugular venous distention	1.05 ± 0.72	0.89 ± 0.66	0.394
Congestive moist rales	0.75 ± 0.58	1.26 ± 0.73	0.009
Gallop rhythm	0.40 ± 0.50	0.58 ± 0.51	0.200
Liver	1.35 ± 0.58	1.37 ± 0.68	0.911
Edema	1.40 ± 0.82	1.11 ± 0.66	0.145

Thus, RV-DCM patients had fewer pronounced signs of LV failure: lower position of the head end of the bed (0.89 ± 0.16 versus 1.58 ± 0.51 , in Group 2, $P = 0.000$) and less congestive

moist rales in the lungs (0.75 ± 0.58 versus 1.26 ± 0.73 in Group 2, $P=0.019$). Such indicators as jugular venous distention and edema in Group 1 patients were prevalent but not statistically significant.

ECG analysis revealed most significant differences in the incidence of “dextrogram” and the ratio of RV6/RVmax amplitude. Particularly, right axis deviation was registered in 16(80%) cases among RV-DCM patients and only in 4(10%) cases in DCM patients with BV-HF ($P=0.000$). RV6/RVmax ratio reached 0.87 ± 0.38 and 2.98 ± 1.29 in Group 1 and Group 2, respectively ($P=0.000$). In addition, 8(40%) patients with RV-DCM had T- wave alternans, which was not recorded in Group 2 ($P=0.000$), while a T-wave inversion occurred in 6(30%) and 13(32%) of cases in Group 1 and Group 2, respectively (Table 3). The patient proportion with persistent AF in Group 1 was slightly above that in Group 2; complete LBBB occurred only in DCM patients with BV-HF ($P=0.129$), and incomplete RBBB was significantly more frequent in RV-DCM patients (5 vs. 1, $P=0.04$). The same frequency of first-degree AV block occurred in both groups, but one RV-DCM patient developed a complete AV block that required pacemaker implantation (Table 3).

Table 3.

ECG parameters of DCM patients

Variable	Group 1	Group 2	P	χ^2
Right axis deviation, high R wave in leads V1-V3 and deep S wave in leads V5-V6, n	16	4	0.000	24.258
T wave inversion in leads V1-V3, without RBBB, n	8	-	0.000	Yates' χ^2 12.952
T wave inversion in leads V5- V6 without LBBB, n	6	13	0.578	0.31
Increases of QRS complex duration >110ms in V1-V)	4	5	0.8791	Yates' χ^2 0.023
Permanent form of AF, n	6	8	0.583	0.301
Complete LBBB, n	-	6	0.129	Yates' χ^2 2.305
Incomplete RBBB, n	5	1	0.04	Yates' χ^2 4.219
Complete RBBB, n	3	2	0.518	Yates' χ^2 0.418
Left posterior hemiblock, n	3	-	0.085	Yates' χ^2 2.961
Left anterior hemiblock, n	1	4	0.747	Yates' χ^2 0.104
First-degree AV block, n	3	2	0.518	Yates' χ^2 0.418
Complete AV block, n	1	-	0.780	Yates' χ^2 0.078

A number of significant differences in the linear and volume indicators were revealed by comparative analysis of Echo-CG parameters (Table 4.) Despite the right-sided heart dilation ($RV=50.54 \pm 8.43$ mm, $RA=47.98 \pm 9.92$ mm), the left-sided heart size remained in normal range ($LVEDD=39.9 \pm 9.8$ mm; $LVESD=29.12 \pm 10.15$ mm; $LA=35.75 \pm 12.69$ mm) that is typical for RV-DCM patients. LV myocardial contractility

also remained within normal range ($LVEF=67.09 \pm 10.0\%$). Right and left heart dilatations with a decrease in inotropic myocardial function were noted in DCM patients with BV-HF.

Table 4.

Echo-CG parameters of DCM patients

Parameter	Group 1	Group 2	P
Aortic Root, mm	30.44 ± 3.69	29.11 ± 4.71	0.274
LA dimension, mm	35.75 ± 12.69	47.48 ± 5.49	0.000
IVST, mm	10.75 ± 1.94	9.62 ± 1.64	0.021
LVPWT, mm	10.45 ± 1.74	9.75 ± 1.95	0.180
LVESD, mm	29.12 ± 10.15	63.73 ± 7.80	0.000
LVEDD, mm	39.9 ± 9.8	74.68 ± 7.34	0.000
LVEF, %	67.09 ± 10.01	30.59 ± 7.13	0.000
LVFS, %	19.63 ± 5.59	14.80 ± 3.37	0.000
RVEF, %	37.86 ± 8.17	47.2 ± 9.31	0.000
RV, mm	50.54 ± 8.43	42.78 ± 5.58	0.000
RA, mm	47.98 ± 9.92	42.98 ± 5.57	0.015
LVEDV, ml	82.30 ± 36.79	296.29 ± 68.61	0.000
LVESV, ml	27.90 ± 16.59	200.88 ± 70.65	0.000
LVSV, ml	54.40 ± 24.81	95.42 ± 27.64	0.000
LVM, g	179.45 ± 67.13	349.46 ± 85.04	0.000
Mean PAP, mmHg	37.12 ± 7.77	44.57 ± 10.75	0.008
Comparative characteristics of LV/RV diastolic function			
LV PE, cm/sec	0.73 ± 0.23	0.66 ± 0.16	0.228
LV PA, cm/sec	0.44 ± 0.21	0.39 ± 0.18	0.367
LV PE/PA	1.57 ± 0.13	1.61 ± 0.11	0.243
RV PE, cm/sec	0.79 ± 0.17	0.67 ± 0.14	0.005
RV PA, cm/sec	0.29 ± 0.31	0.39 ± 0.12	0.077
RV PE/PA	2.64 ± 0.23	1.51 ± 0.11	0.000

Almost all DCM patients had pulmonary arterial hypertension syndrome. Mean pulmonary pressure, defined as an indexed rate, was significantly higher in patients with classical DCM. All Group 1 patients had pseudonormal LVDD ($PE/PA=1.57 \pm 0.11$), but RVDD was worse in Group 2 patients compared to Group 1. The dysfunction found in Group 2 patients was the restrictive type, against the pseudonormal type in Group 1 (2.64 ± 0.23 and 1.57 ± 0.11 ; $P=0.000$).

Results of biochemical and immune-enzyme analyses in DCM are shown in Table 5.

Table 5.

Biochemical parameters of DCM patients

Variable	Group 1	Group 2	P
Total bilirubin, mkmol/l	34.83 ± 23.72	24.44 ± 9.01	0.012
-conjugated	14.82 ± 16.21	6.76 ± 3.51	0.02
-unconjugated	23.26 ± 14.58	21.23 ± 5.59	NS
ALT, IU/l	39.55 ± 42.59	44.92 ± 46.78	NS
AST, IU/l	41.09 ± 47.91	37.33 ± 27.84	NS
Plasma albumin, g/dl	2.9 ± 0.6	3.16 ± 0.48	NS
Total cholesterol, mg/dl	113.8 ± 11.7	107.3 ± 8.8	0.026

A change in level of at least one of any of the functional markers of liver damage was identified in 81.8% of cases in RV-DCM patients, while this index was only 42.1% in Group 2 ($P=0.01$). Hyperbilirubinemia was the most typical marker of the liver damage among examined patients, which was recorded in 91% of patients in Group 1 versus 58% of patients in Group 2. The laboratory analysis showed that the total bilirubin level was increased, mainly due to high values of unconjugated bilirubin fraction in both groups. The serum level of conjugated bilirubin was 2 times greater than the normal value only in Group 1 patients. TNF- α and CRP plasma concentrations were determined additionally in 9(41%) and 14(37%) patients of Group 1 and 2, respectively, and also in 10 healthy persons. The serum level of TNF- α was 59.7 ± 1.8 pg/ml and 58.6 ± 1.68 pg/ml in Group 1 and Group 2, respectively, compared to 29.2 ± 1.4 pg/ml in healthy persons ($P < 0.005$ in both cases).

The increased serum CRP level was without significant differences between groups (9.8 ± 1.1 mg/l and 8.9 ± 1.2 mg/l; $P = 0.084$) and more than three times higher than normal values (2.3 ± 0.6 mg/l; $P < 0.05$ compared to Groups 1 and 2).

In the study of patient prognosis, it was noted that 5-year mortality was almost the same in both groups: 12(54.5%) and 18(47.4%) in Group 1 and Group 2 ($\chi^2 = 0.287$; $P = 0.592$). Analysis of the causes of death revealed that fatal pulmonary embolism (PEmb) was a leading cause (50%) in RV-DCM patients (Fig.1).

Fig. 1. Causes of death in DCM patients

This cause of death was established on the basis of acute symptoms and autopsy results of in-patients, as well as an interview with the relatives when they indicated the suddenly developed dyspnea, cyanosis, hemoptysis which preceded death. The unfavorable outcome of sudden death (SD) in 3(25%) patients of Group 1 occurred as a result of progressive decompensation of CHF. PEmb was the cause of death only in 3(16.7%) cases in Group 2 patients (Yates' $\chi^2 = 2.388$; $P = 0.122$ compared to Group 1). The majority of DCM patients with BV-HF (55.5%) died due to HF progression (Yates' $\chi^2 = 1.635$; $P = 0.201$ compared to Group 1). We also analyzed the frequency of patients' readmissions with CHF destabilization and nonfatal thromboembolic complications, such as acute ischemic stroke and PEmb. Thus, the average number of hospitalizations during the observation period was 8.5 ± 3.2 in Group 2 versus 9.3 ± 2.5 in Group 1 ($P > 0.05$). The cases

of nonfatal stroke and PEmb were registered with different frequency, as expected, on the assumption of morphological and hemodynamic features of heart disease in comparison groups of patients. Acute destabilization of CHF with episodes of hemoptysis was registered in 15(68.2%) of cases in RV-CMP. Thromboembolism was verified by X-ray examination in some patients. The incidence of this outcome was 26,3% ($\chi^2 = 10.048$; $P = 0.0015$) in Group 2. Incidents of ischemic stroke or transient ischemic attack episodes were registered only in patients of Group 2 (7/18.4%; Yates' $\chi^2 = 2.97$; $P = 0.08$).

Results and Discussion

It is well known that cardiomyopathy, in particular DCM, is characterized by extremely poor prognosis and a high mortality rate. However, detection of possible differences in the course and outcome of various DCM clinical variants is very important. We focused on gender features of DCM and its clinical variant, RV-DCM, as an initially diagnosed condition. It is a common view that DCM occurs more frequently in men than in women [2,13], which was confirmed by us. However, women significantly prevailed over men (2.7 times) among RV-DCM patients. It is not possible to confirm or deny this gender feature of RV-DCM because of the very limited publications on the subject. Almost all of them are characterized by descriptions of sporadic cases or small clinical studies. For example, a case of idiopathic dilatation and RV dysfunction in a young 16-year-old girl was described by K.Ishikawa et al.[13]. The monitoring results of 14 RV-DCM patients were published by D.Fitchett et al. [14]. At the same time, Indian researchers (Mohan JC et al., 1989) described RV-DCM in 10 patients, and noted the predominance of women [15]. Obviously, this issue can be clarified by a large epidemiological study. History has shown that RV-DCM patients enter cardiology departments very late due to misdiagnosis or a delay in diagnosis because of the lack the LV dysfunction, which is stereotypically associated with DCM. E. Amosova also indicated that most cases of RV-DCM are diagnosed accidentally during patient examination for intercurrent diseases [16]. According to our data, we can assume that the isolated RV failure is clinically more severe than biventricular heart disease. J.Goldstein [17] noted that RV-MI may result in severe hemodynamic compromise associated with a poor clinical outcome. The ischemic, dyskinetic right ventricular free wall exerts mechanically disadvantageous effects on biventricular performance. Depressed RV systolic function leads to a decrease in transpulmonary delivery of left ventricular (LV) preload, resulting in diminished cardiac output. [17]. The first report of concurrent right and left ventricular hypertrophy in response to pressure or volume overload of the right ventricle was made in 1982. In two pressure and volume overload models of canine right ventricular hypertrophy, D.F. Larson et al. have demonstrated significant hypertrophy of both the left and the right ventricles. The extent of hypertrophy was correlated positively to the extent of the increase in plasma epinephrine in both volume and pressure overload models [18].

Several studies of HF of ischemic etiology have shown

that RV function is related to exercise tolerance and determines the severity of the patient's condition more than LV function, and that increasing RVEF is the measure of the effectiveness of revascularization in CHD patients with congestive HF [19-21]. According to G. Karatasakis et al. [22], in patients with advanced heart failure, preserved RV function as indicated by an echocardiographically derived RV shortening $>1.25\text{cm}$ was a strong predictor of survival.

In addition, we would like to mention the prevalence of some cases of AF in RV-DCM patients although the differences were not significant. The revealed features seemingly contradict the common pathogenesis mechanisms according to which the AF occurrence is mainly due to the appearance of abnormal impulses in LA remodeling from the mouths of the pulmonary veins and throughout the tissue. Nevertheless, the superior vena cava is one of the sources of ectopies that can initiate AF, which was determined in a study performed by M.Goya et al. [23]. Gaynor SL et al. [24] showed that in dogs with chronic RV pressure overload, RV systolic function was preserved, but diastolic function was impaired. To compensate, RA contractility increased, and the atrium became more distensible to maintain filling of the stiffened ventricle. This compensatory response of the right atrium likely plays an important role in preventing clinical failure in chronic pulmonary hypertension. In DCM, it has been histologically shown that a main cause of AF is fibrosis and reduction of myofibrils precisely, and to a lesser extent, remodeling and genetic contribution [25]. These data apparently explain the prevalence of AF in Group 1 patients.

Despite the right-sided heart dilatation in RV-DCM patients ($RV=50.54\pm 8.43\text{mm}$ and $RA=47.98\pm 9.92\text{mm}$), the size of the left-sided heart remained in the normal range ($LVEDD=39.9\pm 9.8\text{mm}$; $LVESD=29.1\pm 10.1\text{mm}$; $LA=35.75\pm 12.69\text{mm}$). The results indicate intact LV myocardial contractility ($LVEF=51.7\pm 10.1\%$). The expected both right and left heart dilatation with marked reduction of inotropic myocardial function was registered in DCM patients with BV-HF. Polak J. F. et al [21] showed that in patients with depressed left ventricular ejection fraction (less than 40%) and clinically evident congestive heart failure secondary to atherosclerotic coronary artery disease the reduction in right ventricular ejection fraction is a useful index not only for assessing biventricular function, but also for predicting patient outcome. These data were confirmed in other studies [5,10].

Studies of the lesser circulation hemodynamics in 150 patients with dilatation cardiomyopathy and in 15 after heart transplantation [26] revealed secondary postcapillary pulmonary hypertension of more than 60mmHg in cases with dilatation cardiomyopathy with systolic pressure in the pulmonary artery (SPPA) in 30% of patients, transpulmonary gradient (TPG) of more than 15 mm Hg and pulmonary vascular resistance (PVR) or more than 4 Wood's U in 15.3% of patients.

One of the anatomical features of the liver is its sophisticated dual blood supply, which makes it quite resistant to hemodynamic overload. It is established that even a slight increase of RA pressure in patients with heart failure may lead to hyperbilirubinemia [15]. Hepatic dysfunction is

developed at the overpressure on 10mmHg and more in RA and a cardiac index reducing to lower than 1.5l/min/m^2 . In cases of "Cytolytic" syndrome with elevated aminotransferase levels, jaundice (defined as bilirubin higher than 3 mg/dL) is a consequence of persistent and severe hypotension and hypoperfusion, such as acute LV failure, PE, and cardiogenic shock.

The results of our study also demonstrated that an increase in the level of liver enzymes was not so pronounced and widespread. An increase of at least one aminotransferase concentration higher than the norm occurred in 36.3% and 30% in Group 1 and Group 2, respectively. Earlier studies have also shown that an increase in the level of liver aminotransferase does not usually exceed normal values by 2 to 3 times in patients with cardiovascular diseases [16].

Hyperbilirubinemia is caused by the violation of the synthesis, metabolism, transport and excretion of bilirubin. This condition is typical for a sufficiently wide variety of pathological conditions, including hepatitis, toxic lesions, biliary obstruction and hepatocellular dysfunction, and hereditary cholestatic syndromes. A secondary liver damage indicator is hyperbilirubinemia, which is characterized mainly by increasing unconjugated bilirubin. However, our results confirm once again the fact that serious damage to the right-sided heart and severe CHF leads to an increase in the level of conjugated bilirubin; at the same time, the jaundice may not manifest clinically before plasma bilirubin levels increase more than 3 mg/dl. Currently, the clinical forms of liver disease in CHF are determined to be congestive hepatopathy, ischemic hepatitis, and cardiac hepatic fibrosis [19]. Changes of liver function test indicators in our study had moderate severity without development of varied clinical symptoms with rare exceptions (refractory ascites, encephalopathy, hemorrhagic complications), which indicated congestive hepatopathy.

The estimation of liver synthetic function is extremely important for the evaluation of the lesion depth. We identified a moderate hypoalbuminemia in both groups of patients (Table 5). It should be noted that the serum albumin does not change in acute viral hepatitis. Drug-induced liver injury, biliary obstruction, and hypoalbuminemia are more common in congestive genesis of liver lesions, enteropathy, nephropathy, and as a complication of heart failure.

In our work, we prospectively analyzed the prognosis of RV-DCM patients on medical therapy according to the CHF therapy guidelines [27,28]. The 5-year mortality of our patients was about 50%, which is consistent with the 5-year survival of 50% to 70% reported in more recent studies on the prognosis of non-ischemic DCM [29-31]. The explicit prevalence of fatal pulmonary embolisms in RV-DCM patients is an important feature of RV-DCM outcomes. Ribeiro and colleagues were the first to suggest that patient with RV failure may have increased mortality. "RV dysfunction when diagnosis of PE is established is associated with mortality rate" [32].

Conclusion

The results of our investigations during many years of research have shown that the clinical incidence of RV-

DCM was 7.3% among all forms of DCM. The study of life prognosis in patients with 2 forms of DCM showed that 5-year mortality of patients was about 50%. Herewith, we detected the differences in causes of death depending on the type of heart damage, primarily development of fatal PE. Obviously, the results we anticipated have limitations, including lack of an analysis of the treatment regimens. Nevertheless, we believe that this pilot study, as well as a number of other small studies, contributes to the clarification of one of the least studied non-coronarogenic myocardial diseases—isolated RV-DCM.

Competing interests

The authors declare that they have no competing interests.

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