ORIGINAL ARTICLE

Idiopathic dilated cardiomyopathy in children: Prognostic indicators

Çocuklarda idiyopatik dilate kardiyomyopati: Prognostik belirteçler

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ABSTRACT

Objective: Dilated cardiomyopathy (DCM) is a disorder featuring left ventricular dysfunction, heart failure, and a poor prognosis. The etiology is still unclear, despite diagnostic and therapeutic developments. This study was an evaluation of factors affecting the life span of a group of idiopathic DCM patients.

Methods: A total of 79 patients from between October 2005 and October 2017 with a diagnosis of idiopathic DCM were evaluated retrospectively. Demographic characteristics, clinical information, left ventricular function, treatment, and follow-up of the patients were reviewed based on hospital records. Age, gender, parental consanguinity, cardiomegaly on telecardiography, reduced ejection fraction (EF) and shortening fraction (SF), degree of mitral regurgitation, and intracardiac thrombosis were determined to affect prognosis.

Results: The patients were aged 20±60 months, and the male/female ratio was 1.02/1. The patients most frequently presented with heart failure signs and symptoms (n=59, 74.7%). The most common physical examination findings were a murmur (n=53, 67.1%) and tachycardia (n=48, 60.8%). Cardiomegaly was observed on telecardiography in 73.4% of the patients. The EF and SF values were 35.7±12.6% and 17.3±6.5%, respectively. In all, 42 (53.2%) patients had mitral regurgitation of grade 2 or higher. The duration of follow-up was between 1 and 156 months (20±34.9 months). Intracardiac thrombosis was detected in 4 (5.1%) patients. The mortality rate was 36.7%. When the prognostic factors were compared according to survival time, it was determined that survival was reduced in cases of parental consanguinity, low EF, and cardiomegaly.

Conclusion: The most important negative markers affecting the length of survival of DCM patients were parental consanguinity, cardiomegaly detected on telecardiography, and a reduced EF level.

ÖZET

Amaç: Dilate kardiyomiyopati (DKM) sol ventrikül işlevlerinde bozukluk ve kalp yetersizliği ile seyreden ve prognozu kötü olan bir hastalıktır. Tanısal ve terapötik gelişmelere rağmen etiyoloji hala bilinmemektedir. Çalışmamızda idiyopatik DKM tanısıyla izlenen hastalarımızın yaşam süresini etkileyen faktörleri değerlendirmek istedik.

Yöntemler: Ekim 2005–Ekim 2017 tarihleri arasında idiyopatik DKM tanısı konulan 79 hasta geriye dönük olarak değerlendirildi. Hastaların demografik özellikleri, klinik bilgileri, sol ventrikül fonksiyonları, tedavi ve izlemleri hastane kayıtlarından incelendi. Prognoza etki edebilecek faktörlerden; semptomların başlama yaşı, cinsiyet, anne-baba akrabalığı, telekardiyogramda kardiyomegali varlığı, ekokardiyografide azalmış ejeksiyon fraksiyonu (EF), kısalma fraksiyonu (KF), mitral yetersizlik derecesi ve intrakardiyak tromboz gelişimi değerlendirildi.

Bulgular: Hastaların yaşları 20±60 ay, erkek/kız oranı ise 1.02/1 idi. Hastaların en sık kalp yetersizliği semptom ve bulguları (n=59, %74.7) ile başvurduğu görüldü. En sık görülen fizik muayene bulgusu üfürüm (n=53, %67.1) ve taşikardi (n=48, %60.8) idi. Olguların %73.4'ünde telekardiyografide kardiyomegali vardı. Ekokardiyografi değerlendirmesinde EF ve KF değerleri sırasıyla %35.7±1.3 ve %17.3±6.5 iken 42 (%53.2) hastada 2. derece ve üzerinde mitral yetersizlik olduğu görüldü. İzlem süresi 20±38.1 (1–156) ay idi. Dört olguda (%5.1) intrakardiyak tromboz saptandı. Mortalite oranı %36.7 olarak bulundu. Sağ kalım sürelerine göre prognostik faktörlere bakıldığında; anne baba arasında akrabalık olması, EF ve telekardiyografide kardiyomegali olmasının sağkalım süresini anlamlı derecede kısalttığı görüldü.

Sonuç: Anne baba arasında akrabalık olması, tanı anında telekardiyografide kardiyomegali olması ve EF değerlerinin düşük olması DKM hastalarında sağkalım sürelerini etkileyen en önemli belirteçlerdir.

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Dilated cardiomyopathy (DCM) is myocardial disease characterized by dilated left ventricular and systolic dysfunction, often coursing with congestive heart failure. The five-year mortality rate or heart transplantation requirement of these patients was reported as 46%.[1] It is the most common type of cardiomyopathy seen in children. The most common cause of DCM is idiopathic and followed by myocarditis. In the United States, annual the incidence of cardiomyopathy in the 0-18 age group is 1.13 per 100,000, and DCMs consist of half of these cases. In children aged under one year, the expected annual incidence of DCM is 4 per 100,000 children.[2] In epidemiological studies, the incidence in children was found to be 0.57–0.73/100.000.[3,4]

The prognosis of patients is in a wide spectrum ranging between sudden improvement and severe heart failure.[3] Due to the fact that drug treatment is not very effective, one third of the patients lead a mortal course or are candidates for heart transplantation.[4]

The most frequency cause of heart transplantation in children is DCM.[5] Early diagnosis and treatment of DCM prevent or delay the development of heart failure. Intracardiac thrombosis is a common complication and a major cause of morbidity and mortality. The incidence of intracardiac thrombosis was found to be between 4–16% in patients with DCM.[6–9] In this study we wanted to investigate the prognostic factors in patients with DCM.

METHODS

In our study, 79 patients with idiopathic DCM who were followed between October 2005 and October 2017 were evaluated retrospectively. The diagnosis of DCM was based on detection of expansion of ventricles and increase in left ventricular wall thickness along with systolic dysfunction detected in two-dimensional echocardiography in accordance with the recommendation of 2006 American Heart Association.[10] Secondary causes of DCM (infections, arrhythmias, endocrine diseases, neuromuscular diseases, rheumatologic and immunological diseases, nutritional deficiencies, conditions leading to ischemia, toxins and systemic diseases) were excluded.

Age and gender distribution, admission symptoms, physical examination, electrocardiography (ECG), telecardiography, echocardiography findings and prognosis were evaluated.

Abbreviations:

DCM	*Dilated cardiomyopathy*
ECG	*Eşlectrocardiography*
EF	*Ejection fraction*
LVEDD	*Left ventricular end-diastolic diameter*
SF	*Shortening fraction*

The age of onset of symptoms (<2 years and, ≥2 years), gender, consanguinity between parents, presence of cardiomegaly in the telecardiogram (0.60 in the neonatal period, 0.55 in the infancy and 0.50 in children) The effects of ejection fraction (EF) and shortening fraction (CF) measurements, left ventricular end-diastolic diameter (LVEDD), and intracardiac thrombosis on prognosis were investigated. The severity of mitral insufficiency was graded based on the extension of mitral regurgitation jet into left atrium as: Grade 1 <15%; Grade 2, 15–30%; Grade 3, 35–50%, and Grade 4, >50% grade 4 mitral regurgitation.[11] The survival of the patients were evaluated from the time of diagnosis.

Statistical analysis

The statistical analysis of the patients' data was performed with "SPSS, Chicago, IL, USA" program. "Survival (Therneau, 2015) package in R Studio" was used to draw the Kaplan-Meier curve. Mean ± standard deviation (SD) was used as descriptive statistics. P value <0.05 was considered significant. The survival times of the patients were evaluated using Kaplan-Meier method and Cox regression analysis. According to single-variable Cox regression results, variables with p<0.25 were selected as candidates for multiple Cox regression modeling.

RESULTS

The median age of 79 patients diagnosed with dilated cardiomyopathy was 20 months (10 days–198 months) while 40 (50.6%) patients were male and 39 (49.4%) of them were female. Forty-four (55.7%) patients were under, and 35 (44.3%) patients above two years of age Heart failure symptoms and signs were the most common cause of admission in both age groups (n=53, 67.1%). The most common physical examination findings were 2–3/6 degree systolic murmur heard at apical localization (n=53, 67.1%) and tachycardia (n=48, 60.4%). In 10 (12.6%) patients, In 30 (38%) patients consanguinity was observed between the parents including 10 patients whose parents. were first-degree relatives.

Electrocardiographic changes were detected in 39 (49.4%) patients and defined as left ventricular hypertrophy in 27 (69.2%), ST-T changes in 8 (20.5%), and arrhytmia in 4 (%10.3) patients.

Of 4 patients with arrhythmia, 1 had supraventricular tachycardia and 3 had rarely seen ventricular extrasystoles. In 58 (73.4%) patients, cardiomegaly was present in the telecardiography. In echocardiographic examination,values for LVEDD, EF and SF were found as 41.7±1.3 mm, 35.7±1.3% and 17.3±6.5, respectively. Patients with mitral regurgitation were examined in groups of Grades <2. and ≥2. Forty-two (53.2%) had ≥2 Grade, and 37 (46.8%) patients had Grade <2. mitral regurgitation. According to age groups of <2 and >2 years; EF values were 36.9±11.8% vs 34.2±13.5%, and SF values were 17.7±5.9% vs 16.9±7.3%, respectively.

The EF and SF values of the patients at the time of admission and at the end of the follow-up period are shown in Table 1. In 4 (5.1%) patients, intracardiac thrombosis was detected which occurred in 3 patients at the time of their first admission and in 1 patient 2 months after the diagnosis (despite the antiaggregant prophylaxis).

EF and SF values of these patients were 25.5±5.9% and 12.2±2.6, respectively. Intracardiac thrombosis was seen in the left ventricle in four, and in the right ventricle in one patient. Two patients were treated with enoxaparin, one patient with heparin and the other one with tissue plasmin activator. None of the patients had a hematologic disorder that could lead to an arrhythmia or procoagulant condition. Demographic, clinical and echocardiographic features of the patients with thrombosis are shown in Table 2.

Table 1. Echocardiographic findings of the patients at admission, and at the end of the follow-up period

	<2 years (%±SD)		≥2 years (%±SD)		All patients (%±SD)	
	EF	SF	EF	SF	EF	SF
At admission	36.9±11.8	17.7±5.9	34.2±13.5	16.9±7.3	35.7±1.3	17.3±6.5
End of follow-up	50.1±21.1	26.2±12.7	46.5±17.7	24.7±11	48.5±1.9	25.5±1.2

EF: ejection fraction; SF: Shortening fraction.

Table 2. Characteristic features of the patients with thrombosis

	Patient no			
	1	2	3	4
Age at diagnosis (year)	4.5	15	2	11
Gender	Male	Male	Male	Female
Development of thrombosis	At the time of diagnosis	At the time of diagnosis	2 months	At the time of diagnosis
Ejection fraction (%)	20	24	20	34
Shortening fraction (%)	10	12	10	16
Left ventricular end-diastolic diameter (mm)	45	57	51	55
Location of the thrombus	LV (n=2), RV (n=1)	LV	LV	LV
Dimensions of the thrombus (mm)	33x14,15x8, 4x4	35x24	40x25	8x8
Prescribed treatment	Heparin	Enoxaparinrine	t-PA	Enoxaparinrine
Duration of treatment (day)	11	30	1	14
Resolution	Complete	Part (4x4)	Died	Complete
Embolism	+	–	–	–

LV: Left ventricle; RV: Right ventricle; tPA: Plasminogen activator tissue plasminogen activator.

Table 3. Distribution of survived, and exited patients

	Number of survived patients (%)	Number of exited patients (%)
Age		
<2 years	30 (68.2)	14 (31.8)
≥2 years	20 (57.1)	15 (42.9)
Gender		
Female	23 (59)	16 (41)
Male	27 (67.5)	13 (32.5)
Consanguinity		
No	37 (75.5)	12 (24.5)
Yes	13 (43.3)	17 (56.7)
Electrographic changes		
No	28 (70)	12 (30)
Yes	22 (56.5)	17 (43.5)
Cardiomegaly detected on telecaridograms		
No	19 (90.5)	2 (9.5)
Yes	31 (53.4)	27 (46.6)
Intracardiac thrombosis		
No	47 (62.7)	28 (37.3)
Yes	3 (75)	1 (25)
Mitral regurgitation		
Grade ≥2	23 (34.5)	19 (65.5)
Grade <2	27 (65.5)	10 (34.5)

The median follow-up period was 20 months (1–156) and 29 patients (36.7%) died during follow-up. Four (5.06%) patients underwent heart transplantation.

Fourteen patients (31.8%) aged <2 years and 15 (42.9%) patients aged ≥2 years exited. The mortality rate was found to be higher in female patients (n=16, 41%) than in male patients (n=13, 32.5%). Seventeen (56.7%) patients with and 12 (24.5%) patients without consanguineous parents exited. Twenty-seven (46.6%) patients with 2 (9.5%) patients without cardiomegaly as detected on telecardiography did not survive.

The mortality rate was 65.5% (n=19) in patients with mitral regurgitation at grade 2 and above (Table 3). EF values of living and deceased patients were 38.6±12.6% and 30.7±11.1, respectively, while SF values were 18.6±6.7% and 15±5.6, respectively.

Thirty-seven patients (46.8%) with Grade 1 and 42 (53.2%) cases with Grade ≥2 had mitral regurgitation. Of the 4 patients with thrombosis, 2 had Grade 2 and 1 had Grade I mitral regurgitation, while the fourth patient had no mitral regurgitation. The survival times of the patients were evaluated with Cox regression method and Kaplan-Meier method. Variables within a significance level of $p<0.25$ in univariate Cox regression analysis were selected as candidate risk factors considering that they may have significance in multivariate Cox proportional hazard regression analysis.

A retrospective elimination process was performed with multiple analysis. Three variables were left; Parental consanguinity ($p=0.003$), cardiomegaly in telecardiography ($p=0.085$) and low EF value ($p=0.088$) were found to be the most important factors in determining survival (Table 4). The presence of cardiomegaly in the telecardiography and low EF were clinically significant, although not statistically significant, on survival. Because the risk of death was 3.67 times higher in patients with cardiomegaly, each 1 unit EF decreased the risk of death by 1.03 times. The survival curves of the patients are shown in Figure 1–3.

Categorical risk factors were evaluated by Kaplan-Meier survival analysis using Log-Rank test (Table 5). Numerical variables were evaluated by the proportional hazard regression method of univariate Cox analysis (Table 6).

DISCUSSION

The most frequent symptoms and signs of dilated cardiomyopathy are dyspnea, exercise intolerance, syncope, tachypnea, tachycardia and hepatomegaly.[12] Talierci et al.[13] found that heart failure was the most common (92%) finding in 24 patients with a mean age of two years.

In a study by Oh et al.[14] it was seen that the patients were admitted most frequently (67.1%) with signs and symptoms of heart failure. We found that 73.4% of our patients had cardiomegaly as detected in telecardiography.

In many studies, patients with a family history of CDM have been evaluated and it has been shown that patients have one or more relatives with CDM.[15–18] Towbin et al.[1] reported the best 5-year survival rate of 94% in patients with family history of CDM.

Table 4. Effects of risk factors on prognosis

	Variable	B	SE	Z	P	HR	95% CI	
							Lower	Upper
Step 1	Age	0.399	0.412	0.936	0.333	1.49	0.664	3.343
	Gender	0.14	0.402	0.122	0.727	1.15	0.524	2.527
	Consanguinity	0.943	0.442	4.988	0.026	2.568	1.122	5.87
	Cardiomegaly in telecardiogram	1.287	0.782	2.709	0.1	3.62	0.782	16.75
	Grade of mitral regurgitation	0.105	0.551	0.036	0.849	1.111	0.377	3.269
	Intracardiac thrombus	-0.124	1.062	0.014	0.907	0.884	0.11	7.078
	Left venticular enddiastolic diameter	-0.007	0.017	0.184	0.668	0.993	0.96	1.026
	Ejection fraction	-0.021	0.065	0.102	0.749	0.979	0.862	1.113
	Shortening fraction	-0.012	0.126	0.01	0.921	0.988	0.772	1.263
Step 2	Age	0.395	0.41	0.925	0.336	1.484	0.664	3.316
	Gender	0.146	0.397	0.134	0.714	1.157	0.531	2.521
	Consanguinity	0.926	0.383	5.85	0.016	2.523	1.192	5.34
	Cardiomegaly in telecardiogram	1.302	0.766	2.886	0.089	3.677	0.819	16.51
	Grade of mitral regurgitation	0.094	0.538	0.03	0.862	1.098	0.383	3.15
	Intracardiac thrombus	-0.112	1.055	0.011	0.915	0.894	0.113	7.07
	Left venticular enddiastolic diameter	-0.007	0.017	0.176	0.675	0.993	0.961	1.026
	Ejection fraction	-0.027	0.021	1.719	0.19	0.973	0.973	1.013
Step 3	Age	0.39	0.41	0.913	0.339	1.479	0.663	3.3
	Gender	0.143	0.397	0.131	0.718	1.154	0.530	2.51
	Consanguinity	0.927	0.382	5.885	0.015	2.528	1.195	5.35
	Cardiomegaly in telecardiogram	1.3	0.766	2.879	0.09	3.67	0.817	16.46
	Grade of mitral regurgitation	0.08	0.523	0.024	0.878	1.084	0.388	3.02
	Left venticular enddiastolic diameter	-0.007	0.017	0.178	0.682	0.993	0.961	1.026
	Ejection fraction	-0.027	0.02	1.855	0.173	0.973	0.935	1.012
Step 4	Age	0.39	0.41	0.91	0.34	1.478	0.663	3.29
	Gender	0.133	0.391	0.116	0.734	1.142	0.531	2.458
	Consanguinity	0.934	0.379	6.062	0.014	2.545	1.21	5.355
	Cardiomegaly in telecardiogram	1.302	0.765	2.897	0.089	3.677	0.821	16.47
	Left venticular enddiastolic diameter	-0.006	0.016	0.146	0.703	0.994	0.963	1.026
	Ejection fraction	-0.029	0.017	3.022	0.082	0.971	0.940	1.004
Step 5	Age	0.388	0.412	0.888	0.346	1.474	0.658	3.304
	Consanguinity	0.938	0.379	6.117	0.013	2.555	1.215	5.375
	Cardiomegaly in telecardiogram	1.266	0.758	2.789	0.095	3.547	0.803	15.667
	Left venticular enddiastolic diameter	-0.006	0.016	0.119	0.730	0.994	0.963	1.027
	Ejection fraction	-0.028	0.016	2.922	0.087	0.972	0.942	1.004
Step 6	Age	0.329	0.375	0.769	0.38	1.389	0.666	2.896
	Consanguinity	0.942	0.379	6.163	0.013	2.564	1.219	5.393
	Cardiomegaly in telecardiogram	1.272	0.757	2.825	0.093	3.567	0.810	15.716
	Ejection fraction	-0.028	0.017	2.776	0.096	0.973	0.942	1.005
Step 7	Consanguinity	0.926	0.379	5.978	0.014	2.523	1.202	5.299
	Cardiomegaly in telecardiogram	1.300	0.755	2.962	0.085	3.669	0.835	16.126
	Ejection fraction	-0.028	0.017	2.904	0.088	0.972	0.941	1.004

In our study, none of our patients had a family history of DCM. In our country, consanguineous marriages are still an important problem. The rate of kinship of parents of our patients was found to be 38 percent. Family members were evaluated echocardiographically in our study and no history of heart disease was reported.

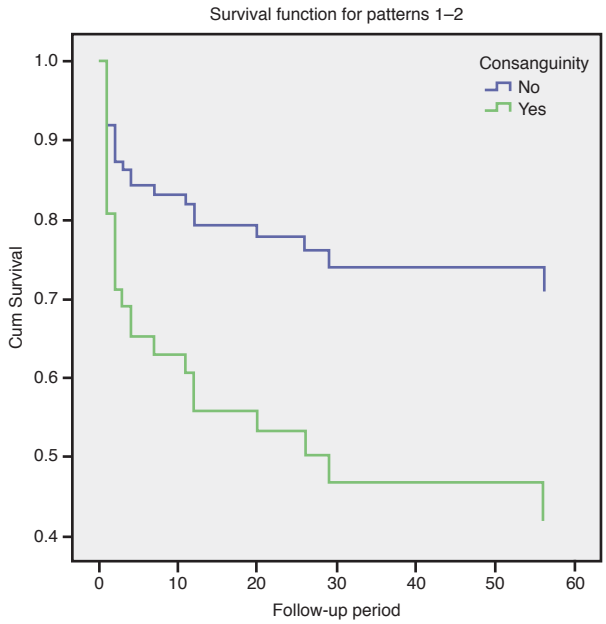

Figure 1. The effect of parental consanguinity on survival times.

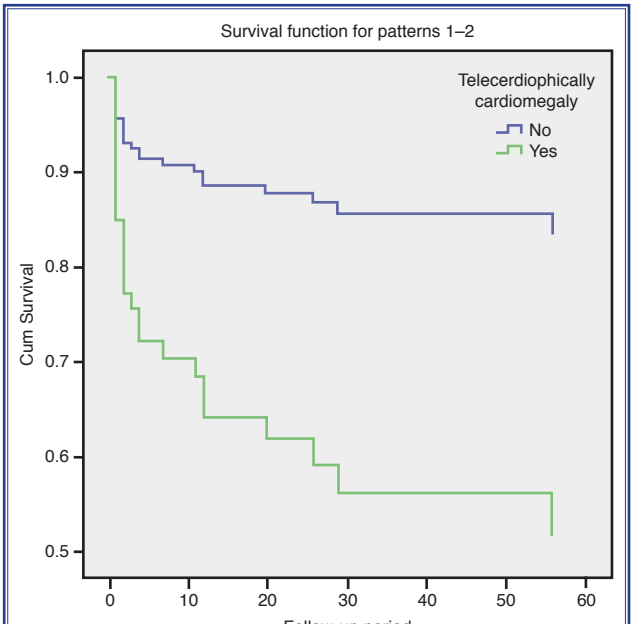

Figure 2. The impact of telecerdiophically detected cardiomegaly on survival.

However, in our country where the rate of consanguineous marriages is high, we think that family members should have detailed cardiological/echocardiographic examination and genetic studies if necessary.

The incidence of intracardiac thrombosis in children with dilated cardiomyopathy has been reported as 4–16% in various studies.[7,19,20] We found its incidence as 5.1% in our study population.

The incidence of thrombosis has been shown to increase up to 43–57% in autopsy studies.[20–22]

When the effect of age on prognosis is evaluated, there are different results in the literature. Carvalho et al.[23] reported a worse prognosis in patients over two years of age. In the study performed by Kim et al.,[24] age was not found to have an effect on prognosis. In our study, we found that age did not have any effect on mortality.

The relationship between gender of the patients and prognosis in patients with CDM has been evaluated, and in some studies female, in others, male gender was reported to be associated with poor prognosis.[15,25] In our study we have not detected any effect of gender on prognosis.

Cardiomegaly and pulmonary edema are the most common findings in telecardiogram sof patients with dilated cardiomyopathy.[26] Increased cardiothoracic ratio is predictive of mortality.[27] In our study, it was found to be effective on mortality.

Taliercio et al.[13] reported lack of any relationship between left ventricular function and prognosis. Zecchin et al.[28] reported that patients with a longer LVEDD, who did not use beta-blockers and had a EF of less than 30% had a poor prognosis. McMahon et al.[29] reported a poorer prognosis for patients with

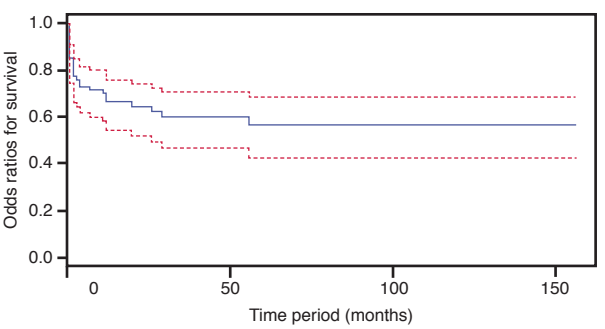

Figure 3. Survival curves of the patients.

Table 5. Evaluation of categorical risk factors with Kaplan-Meier survival analysis using Log-rank test

	Median survival times (month)	CI (95%)		Log-Rank	
		Lower	Upper	Chi	*p*
Age					
<2	92.3	72.6	112	1.58	0.209
>2	76.8	47.1	106.5		
Gender					
Female	87	61.3	112.6	0.56	0.45
Male	74.5	56.5	92.5		
Consanguinity					
No	115.7	96.6	134.8	10.85	0.001
Yes	41.2	18.4	64		

Table 6. Evaluation of numerical variables presumptively effectivbe on survival rates using univariate Cox proportional hazars regression analysis

	Hazard Ratio	CI (95%)		Wald	*p*
		Lower	Upper		
Age	1.56	0.75	3.24	1.43	0.23
Left ventricular enddiastolic diameter	1.01	0.98	1.04	0.38	0.537
Ejection fraction	0.96	0.93	0.99	6.73	0.009
Shortening fraction	0.93	0.88	0.99	5.05	0.025

thicker left ventricular posterior wall, longer LVEDD and low EF.

We found that EF and SF values of the deceased patients were significantly lower. However, we found no significant difference in the mortality rates in patients with a mortal course and a wider LVEDD compared with the survivors.

Another factor effective on prognosis is the development of intracardiac thrombosis.

In our study, we found that the prognosis of our patients with intracardiac thrombosis was not statistically worse. We thought that this result may be related to our limited number of patients. In the etiology of intracardiac thrombosis in children lower cardiac output, arrhythmia, and hematological diseases which result in hypercapulopathy are shown as three main etiologic factors.[6,19,30]

Our patients did not have an etiologic factor or arrhythmia that could cause hypercoagulopathy. Therefore, it was thought that thrombosis was caused by low cardiac output, that is, stasis in blood flow. In studies performed especially in adult patients, it was observed that development of thrombosis was less frequent in patients with mitral regurgitation which was explained by the prevention of blood stasis in mitral regurgitation.[8,31,32] However, in another study, such a relationship could not be found.[19] Similarly, in our study, development of thrombosis was observed in our patients despite the presence of second and third degree mitral insufficiency. Any relationship between regurgitation and thrombosis could not be found. In pediatric patients with DCM, intracardiac thrombus is frequently localized in the left ventricle as was the case with our patients. Though rarely development of thrombosis has been reported in other cardiac chambers.[6,7,19,30]

Treatment of Intracardiac thrombosis is treated via medical [thrombolytic, anticoagulant] or surgical methods.[33,34]

There is no precise information about the choice of treatment options in children. Medical treatment has a tendency to treat small thromboembolism. However, as is shown, morphology of the thrombi has not an effective factor on the risk of embolism.[30] If a patient with CDM has SF less than 20%, then it is recommended to start intensive anticoagulant therapy if the patient has a history of stroke and/or if an intracardiac thrombus is observed.[7,19]

Günthard et al.[6] suggested initiation of intravenous heparin or oral anticoagulant therapy when an intracardiac thrombus is observed. CDM patients with large thrombi need thrombolytic therapy. Thrombolytic therapy was started in one of our patients because thrombus was very large (40x25 mm).

In patients with dilated cardiomyopathy, the prognosis is poor despite medical treatment. Heart transplantation should be considered in patients with multiple organ failure and require treatment with mechanical ventilator despite medical treatment.[26] In our study, heart transplantation was performed in four (5%) patients. The mortality rate has been reported to be between 11.5 and 26.9% in the studies performed. In our study, the mortality rate in our patients was found to be 36.7 percent.

In conclusion, the main factors affecting mortality and five-year survival in patients with DCM have been found as kinship marriages, detection of cardiomegaly in telecardiograms at the time of diagnosis, and lower EF and SF values, and higher grades of mitral regurgitation in echocardiography.

Ethics Committee Approval: Approval was obtained from the Ethics Committee of Yenimahalle Training and Research Hospital (decisionj no: 2017/10/09 File no: 2017/57).

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REFERENCES

1. Towbin JA, Lowe AM, Colan SD, Sleeper LA, Orav EJ, Clunie S, et al. Incidence, causes, and outcomes of dilated cardiomyopathy in children. JAMA 2006;296:1867–76.
2. Elliott P, Andersson B, Arbustini E, Bilinska Z, Cecchi F, Charron P, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J 2008;29:270–6.
3. Nugent AW, Daubeney PE, Chondros P, Carlin JB, Cheung M, Wilkinson LC, et al; National Australian Childhood Cardiomyopathy Study. The epidemiology of childhood cardiomyopathy in Australia. N Engl J Med 2003;348:1639–46.
4. Strauss A, Lock JE. Pediatric cardiomyopathy-a long way to go. N Engl J Med 2003;348:1703–5.
5. Kirk R, Dipchand AI, Rosenthal DN, Addonizio L, Burch M, Chrisant M, et al. The International Society for Heart and Lung Transplantation Guidelines for the management of pediatric heart failure: Executive summary. J Heart Lung Transplant 2014;33:888–909.
6. Günthard J, Stocker F, Bolz D, Jäggi E, Ghisla R, Oberhänsli I, et al. Dilated cardiomyopathy and thrombo-embolism. Eur J Pediatr 1997;156:3–6.
7. Choi SH, Jeong SI, Yang JH, Kang IS, Jun TG, Lee HJ, et al. A single-center experience with intracardiac thrombosis in children with dilated cardiomyopathy. Pediatr Cardiol 2010;31:264–9.
8. Falk RH, Foster E, Coats MH. Ventricular thrombi and thromboembolism in dilated cardiomyopathy: a prospective follow-up study. Am Heart J 1992;123:136–42.
9. Irdem A, Başpınar O, Kervancıoğlu M, Kılınç M. Intracardiac thrombus in children with dilated cardiomyopathy. Turk Kardiyol Dern Ars 2014;42:161–7.
10. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, et al.; American Heart Association; Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; Council on Epidemiology and Prevention. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. Circulation 2006;113:1807–16.
11. Apostolakis EE, Baikoussis NG. Methods of estimation of mitral valve regurgitation for the cardiac surgeon. J Cardiothorac Surg 2009;4:34.
12. Press S, Lipkind RS. Acute myocarditis in infants. Initial presentation. Clin Pediatr (Phila) 1990;29:73–6.
13. Taliercio CP, Seward JB, Driscoll DJ, Fisher LD, Gersh BJ, Tajik AJ. Idiopathic dilated cardiomyopathy in the young: clinical profile and natural history. J Am Coll Cardiol 1985;6:1126–31.
14. Oh JH, Hong YM, Choi JY, Kim SJ, Jung JW, Sohn S, et al.

Idiopathic cardiomyopathies in Korean children. - 9-Year Korean Multicenter Study. Circ J 2011;75:2228–34.
15. Kumar RK. A practical approach for the diagnosis and management of dilated cardiomyopathy. Indian J Pediatr 2002;69:341–50.
16. Grünig E, Tasman JA, Kücherer H, Franz W, Kübler W, Katus HA. Frequency and phenotypes of familial dilated cardiomyopathy. J Am Coll Cardiol 1998;31:186–94.
17. Mahon NG, Murphy RT, MacRae CA, Caforio AL, Elliott PM, McKenna WJ. Echocardiographic evaluation in asymptomatic relatives of patients with dilated cardiomyopathy reveals preclinical disease. Ann Intern Med 2005;143:108–15.
18. Rusconi P, Wilkinson JD, Sleeper LA, Lu M, Cox GF, Towbin JA, et al.; Pediatric Cardiomyopathy Registry Investigators. Differences in Presentation and Outcomes Between Children With Familial Dilated Cardiomyopathy and Children With Idiopathic Dilated Cardiomyopathy: A Report From the Pediatric Cardiomyopathy Registry Study Group. Circ Heart Fail 2017;10. pii: e002637.
19. McCrindle BW, Karamlou T, Wong H, Gangam N, Trivedi KR, Lee KJ, et al. Presentation, management and outcomes of thrombosis for children with cardiomyopathy. Can J Cardiol 2006;22:685–90.
20. Akagi T, Benson LN, Lightfoot NE, Chin K, Wilson G, Freedom RM. Natural history of dilated cardiomyopathy in children. Am Heart J 1991;121:1502–6.
21. Chen SC, Nouri S, Balfour I, Jureidini S, Appleton RS. Clinical profile of congestive cardiomyopathy in children. J Am Coll Cardiol 1990;15:189–93.
22. Harris LC, Rodin AE, Nghiem QX. Idiopathic, nonobstructive cardiomyopathy in children. Am J Cardiol 1968;21:153–65.
23. Carvalho JS, Silva CM, Shinebourne EA, Redington AN. Prognostic value of posterior wall thickness in childhood dilated cardiomyopathy and myocarditis. Eur Heart J 1996;17:1233–8.
24. Kim IS, Izawa H, Sobue T, Ishihara H, Somura F, Nishizawa T, et al. Prognostic value of mechanical efficiency in ambulatory patients with idiopathic dilated cardiomyopathy in sinus rhythm. J Am Coll Cardiol 2002;39:1264–8.
25. Wilkinson JD, Landy DC, Colan SD, Towbin JA, Sleeper LA, Orav EJ, et al. The pediatric cardiomyopathy registry and heart failure: key results from the first 15 years. Heart Fail Clin 2010;6:401–13.
26. Jefferies JL, Towbin JA. Dilated cardiomyopathy. Lancet 2010;375:752–62.
27. Manolio TA, Baughman KL, Rodeheffer R, Pearson TA, Bristow JD, Michels VV, et al. Prevalence and etiology of idiopathic dilated cardiomyopathy (summary of a National Heart, Lung, and Blood Institute workshop. Am J Cardiol 1992;69:1458–66.
28. Zecchin M, Lenarda AD, Bonin M, Mazzone C, Zanchi C, Di Chiara C, et al. Incidence and predictors of sudden cardiac death during long-term follow-up in patients with dilated cardiomyopathy on optimal medical therapy. Ital Heart J 2001;2:213–21.
29. McMahon CJ, Nagueh SF, Eapen RS, Dreyer WJ, Finkelshtyn I, Cao X, et al. Echocardiographic predictors of adverse clinical events in children with dilated cardiomyopathy: a prospective clinical study. Heart 2004;90:908–15.
30. John JB, Cron SG, Kung GC, Mott AR. Intracardiac thrombi in pediatric patients: presentation profiles and clinical outcomes. Pediatr Cardiol 2007;28:213–20.
31. Kalaria VG, Passannante MR, Shah T, Modi K, Weisse AB. Effect of mitral regurgitation on left ventricular thrombus formation in dilated cardiomyopathy. Am Heart J 1998;135:215–20.
32. Ozdemir N, Kaymaz C, Daglar E, Karakaya O, Akçay M, Ozkan M. Severe mitral regurgitation may prevent mural thrombus formation within the left ventricle with systolic dysfunction. Jpn Heart J 2002;43:495–503.
33. Aspesberro F, Beghetti M, Oberhänsli I, Ozsahin H, Humbert J, Rimensberger PC. Local low-dose urokinase treatment of acquired intracardiac thrombi in preterm infants. Eur J Pediatr 1999;158:698–701.
34. Paç FA, Cağdaş DN. Treatment of massive cardiac thrombi in a patient with protein C and protein S deficiency. Blood Coagul Fibrinolysis 2007;18:699–702.
35. Bilgiç A, Ozbarlas N, Ozkutlu S, Ozer S, Ozme S. Cardiomyopathies in children. Clinical, epidemiological and prognostic evaluation. Jpn Heart J 1990;31:789–97.
36. Burch M, Siddiqi SA, Celermajer DS, Scott C, Bull C, Deanfield JE. Dilated cardiomyopathy in children: determinants of outcome. Br Heart J 1994;72:246–50.

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