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ORIGINAL ARTICLE

Retrospective Study

Contemporary characteristics and outcomes of adults with familial dilated cardiomyopathy listed for heart transplantation

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Abstract

BACKGROUND

Familial dilated cardiomyopathy (FDCM) account for 20%-30% of non-ischemic cardiomyopathies (NICM). Previous published data showed that some patients with FDCM tend to have rapidly progressive disease; however, five-year mortality was not significantly different in the familial and non-familial forms of NICM with optimal medical therapy.

AIM

To better define the characteristics and clinical outcomes of FDCM patients listed for heart transplantation (HT).

METHODS

We queried the United Network for Organ Sharing Registry to identify FDCM patients listed for HT between January 2008 and September 2015 and compared them to NICM and ischemic cardiomyopathy (ICM) patients. We included all patients ≥ 18 years old and we separated patients to three groups: FDCM, NICM and ICM. Chi-square test was used to compare between categorical variables, the t-test was used to compare between continues variables, and Cox-proportional hazards model was used to perform time-dependent survival analyses.

RESULTS

Of the 24809 adults listed for HT, we identified 677 patients (2.7%) with the diagnosis of FDCM. Compared to patients with NICM and ICM, FDCM patients were younger (FDCM 43.9 ± 13.5 *vs* NICM 50.9 ± 12.3, $P < 0.001$, *vs* ICM 58.5 ± 8.1, $P < 0.001$), more frequently listed as status 2 (FDCM 35.2% *vs* NICM 26.5%, $P < 0.001$), with significantly lower left ventricular assist device (LVAD) utilization (FDCM 18.4% *vs* NICM 25.1%, $P < 0.001$; *vs* ICM 25.6%, $P < 0.001$), but higher use of total artificial heart (FDCM 1.3% *vs* NICM 0.6%, $P = 0.039$; *vs* ICM 0.4%, $P =$

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0.002). Additionally, patients with FDCM were less frequently delisted for clinical deterioration or death and more likely to be transplanted compared to those with NICM [hazard ratio (HR): 0.617, 95% confidence interval (CI): 0.47-0.81; HR: 1.25, 95%CI: 1.14-1.37, respectively], and ICM (HR: 0.5, 95%CI: 0.38-0.66; HR: 1.18, 95%CI: 1.08-1.3, respectively). There was more frequent rejection among patients with FDCM (FDCM 11.4% *vs* NICM 9.8%, $P = 0.28$; *vs* ICM 8.4%, $P = 0.034$). One, three, and five post-transplant survival of patients with FDCM (91%, 88% and 80%) was similar to those with NICM (91%, 84%, 79%, $P = 0.225$), but superior to those with ICM (89%, 82%, 75%, $P = 0.008$), respectively.

CONCLUSION
End-stage FDCM patients are more likely to be transplanted, more likely to have early rejection, and have similar or higher survival than patients with other cardiomyopathies.

Key words: Familial dilated cardiomyopathy; End-stage heart failure; Wait list; Transplant; Outcomes

Core tip: Familial dilated cardiomyopathy (FDCM) can lead to end-stage heart failure requiring heart transplantation (HT). There is little contemporary information on progression, circulatory mechanical support use, and HT outcomes of these patients. We aimed to define the characteristics and outcomes of FDCM patients and to compare FDCM to non-ischemic cardiomyopathy (NICM) and ischemic cardiomyopathy (ICM) patients listed for HT. FDCM patients were younger, more frequently listed as status 2, and more likely to be transplanted. There was more frequent rejection among patients with FDCM compared to ICM. Post-transplant survival of FDCM patients was similar to NICM, but superior to ICM patients.

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INTRODUCTION

Familial dilated cardiomyopathy (FDCM) account for 20%-30% of non-ischemic cardiomyopathies (NICM)[1-3]. They are most often inherited in a Mendelian autosomal dominant fashion, although autosomal recessive or X-linked transmission exists[4]. Therefore, first-degree relatives have a higher risk of developing the disease[5]. In the United States, around 26% of patients listed for heart transplantation (HT) in the United Network for Organ Sharing (UNOS) Registry are diagnosed with FDCM[6]. Most previous outcome studies of NICM have not studied FDCM as a separate entity[2,7], perhaps because of the challenge in identifying these patients[2]. To make the diagnosis of FDCM, patients should have two or more affected relatives with NICM or a relative of a NICM patient with unexplained sudden death before the age of 35 years[8,9]. Previous published data showed that some patients with FDCM tend to have rapidly progressive disease[1,2], however, five-year mortality was not significantly different in the familial and non-familial forms of NICM with optimal medical therapy[2,3]. Similarly, mechanical circulatory support (MCS) utilization and HT outcomes have not well studied in FDCM patients and most available data are derived from relatively small cohorts and case reports. In this study, we used a large, contemporary, nationwide database to investigate the clinical characteristics, natural history, MCS use, and HT outcomes of patients with end-stage heart failure due to FDCM.

MATERIALS AND METHODS

Data source

We used the thoracic transplantation files from the UNOS Registry contracted with the Health Resources and Services Administration. UNOS includes transplantation information on listed patients in all centers across the United States. Data are collected at different time points: at listing, before transplantation, and continually after transplantation. The listing center is responsible for providing the data. Data is used to match patients with donors, for administrative purposes, and for research reporting. The UNOS registry includes data on patient demographics, cause of cardiomyopathy, implanted devices, causes of removal from wait list, hemodynamics, comorbid conditions, listing status, laboratory tests, donor demographics, laboratory and other testing, post-transplantation complications [rejection, infection, kidney failure, length of stay (LOS)], vital status, and cause of death. The registry is continuously audited with strict quality control[10]. Data included in the UNOS are extracted from the transplant candidate registration form, which is filled at time of transplantation; and transplant recipient follow-up form, which is filled at follow-up. At the time of analysis, the database included 99177 patients listed for HT (1985-2015).

Patient population

We included adults (≥ 18 years old), listed for HT with a diagnosis of idiopathic dilated cardiomyopathy "Dilated myopathy: idiopathic", FDCM "Dilated Myopathy: Familial" and ischemic cardiomyopathy (ICM) "Dilated Myopathy: Ischemic", between January 1st, 2008 to September 30th, 2015. We separated patients to three groups: FDCM, NICM and ICM and compared them. Additional cases were identified in the diagnosis free text variable. We compared their baseline characteristics, MCS utilization, and post-transplant outcomes to patients with the diagnosis of ICM and NICM.

Statistical analyses

All analyses were performed using Statistical Package for Social Sciences (SPSS, version 19.0; SPSS Inc, Chicago, IL). The primary outcomes of this study were waitlist mortality/delisting for clinical deterioration, and post-transplantation mortality among patients who undergo transplantation. Secondary outcomes were as follows: delisting due to improvement, transplant, post-transplantation stroke, post-transplantation permanent pacemaker implantation, post-transplantation acute rejection, post-transplantation dialysis, and LOS for index transplant hospitalization.

Categorical variables were presented as numbers and percentages and were compared using Pearson χ^2 test. Continuous variables were presented as means and standard deviations and were compared with Student *t*-test and. Survival analyses were done using Kaplan-Meier method with log-rank test and adjusted survival using Cox-proportional-hazard model. Variables that were significant in univariable models ($P < 0.05$) were included in the multivariable model. All tests were two sided. $P < 0.05$ was considered statistically significant. No assumptions were used for missing data. Institutional review board approval was not required because only deidentified data sets were used for this analysis. The statistical review of the study was performed by a biomedical statistician.

RESULTS

Demographic characteristics

Of the 24809 adults listed for HT between January 2008 and September 2015, we identified 677 patients (2.7%) with the diagnosis of FDCM, and compared them with 8416 patients (33.9%) with NICM, and 8301 (33.5%) patients with ICM patients (Table 1).

Patients with FDCM were younger (mean age: 43.9 ± 13.5 *vs* NICM 50.9 ± 12.3, $P < 0.001$; *vs* ICM 58.5 ± 8.1; $P < 0.001$) and less predominantly men (FDCM 65.6% *vs* NICM 72.6%, $P < 0.001$; *vs* ICM 86.9%, $P < 0.001$). FDCM patients were more often listed as a status 2 (FDCM 35.2% *vs* NICM 26.5%, $P < 0.001$ *vs*, ICM 34.1%, $P = 0.956$), had significantly less left ventricular assist device (LVAD) use (FDCM 18.4% *vs* NICM 25.1%, $P < 0.001$; *vs* ICM 25.6%, $P < 0.001$) but more use of total artificial heart (TAH) (FDCM 1.3% *vs* NICM 0.6%, $P = 0.039$; *vs* ICM 0.4%, $P = 0.002$), had lower creatinine (FDCM 1.3 ± 0.7 *vs* NICM 1.4 ± 1.0, $P = 0.008$; *vs* ICM 1.4 ± 0.9, $P < 0.001$), had higher albumin (FDCM 3.8 ± 0.6 *vs* NICM 3.7 ± 0.7, $P = 0.001$; *vs* ICM 3.7 ± 0.7, $P = 0.001$), had lower pulmonary artery systolic pressure (FDCM 42.7 ± 13.2 *vs* NICM 44.73 ± 13.9, $P = 0.004$; *vs* ICM 44.8 ± 15.2; $P = 0.001$) , and lower cardiac output (FDCM 4.1 ± 1.3 *vs* NICM 4.3 ± 1.4, $P = 0.011$; *vs* ICM 4.5 ± 1.3, $P < 0.001$) (Table 1).

Table 1 Baseline characteristics by etiology *n* (%)

	NICM (*n* = 8416)	FDCM (*n* = 677)	*P* value	ICM (*n* = 8301)	*P* value
Age at listing	50.9 ± 12.3	43.9 ± 13.5	< 0.001	58.5 ± 8.1	< 0.001
Male gender	6113 (72.6)	444 (65.6)	< 0.001	7212 (86.9)	< 0.001
Ethnicity			< 0.001		< 0.001
White	4609 (54.8)	444 (65.6)		6411 (77.2)	
Black	2776 (33.0)	159 (23.5)		976 (11.8)	
Hispanic	705 (8.4)	54 (8.0)		575 (6.9)	
Asian	223 (2.6)	14 (2.1)		269 (3.2)	
Other or unknown	103 (1.2)	6 (0.9)		70 (0.8)	
Initial status			< 0.001		0.956
1A	1918 (22.8)	147 (21.7)		1823 (22.0)	
1B	4023 (47.8)	273 (40.3)		3415 (41.1)	
2	2227 (26.5)	238 (35.2)		2831 (34.1)	
7	248 (2.9)	19 (2.8)		232 (2.8)	
Therapies					
Inotropes	2947 (35)	233 (34.4)	0.769	2386 (28.7)	0.002
ECMO	52 (0.6)	4 (0.6)	1.0	87 (1.0)	0.320
IABP	395 (4.7)	21 (3.1)	0.056	403 (4.9)	0.038
Mechanical ventilation	141 (1.7)	10 (1.5)	0.875	209 (2.5)	0.118
LVAD	2104 (25.1)	124 (18.4)	< 0.001	2116 (25.6)	< 0.001
BiVAD	153 (1.8)	8 (1.2)	0.288	157 (1.9)	0.233
TAH	50 (0.6)	9 (1.3)	0.039	31 (0.4)	0.002
ICD	6985 (83.5)	562 (83.8)	0.914	6652 (80.9)	0.073
Laboratory values					
Creatinine	1.4 ± 1.0	1.3 ± 0.7	0.008	1.4 ± 0.9	< 0.001
Albumin	3.7 ± 0.7	3.8 ± 0.6	0.001	3.7 ± 0.7	0.001
Bilirubin	1.1 ± 1.8	1.1 ± 1.0	0.578	1.0 ± 2.0	0.540
PRA class I	7.1 ± 18.6	7.2 ± 18.5	0.892	5.7 ± 16.2	0.084
PRA class II	4.8 ± 15.7	5.2 ± 16.2	0.645	3.4 ± 12.6	0.012
Hemodynamics					
PA systolic pressure (mmHg)	44.3 ± 13.9	42.7 ± 13.2	0.004	44.8 ± 15.2	0.001
PA diastolic pressure (mmHg)	22.1 ± 8.9	21.7 ± 8.7	0.266	20.8 ± 8.6	0.013
PA mean pressure (mmHg)	30.5 ± 10.2	29.7 ± 9.8	0.052	29.8 ± 10.5	0.821
PCWP (mmHg)	20.5 ± 9.0	20.4 ± 8.5	0.735	19.7 ± 8.9	0.081
CO (L/min)	4.3 ± 1.4	4.1 ± 1.3	0.011	4.5 ± 1.3	< 0.001

NICM: Non-ischemic cardiomyopathy; FDCM: Familial dilated cardiomyopathy; ICM: Ischemic cardiomyopathy; UNOS: United network for organ sharing; ECMO: Extracoroporeal membrane oxygenation; IABP: Intraaortic balloon pump; VAD: Ventricular assist device; LVAD: Left ventricular assist device; BiVAD: Biventricular assist device; TAH: Total artificial heart; ICD: Implantable cardioverter-defibrillator; PA: Pulmonary artery; PCWP: Pulmonary capillary wedge pressure; CO: Cardiac output.

Wait list outcome

Of 677 FDCM patients, 33 patients (4.8%) died while waiting HT, 7 patients (1%) were delisted for improvement, 20 patients (2.9%) were delisted for deterioration, 470 patients (69%) were transplanted, 3 patients (0.4%) refused transplantation, and 13 patients (1.9%) transferred to another center. Causes of Death in FDCM patients were: multiple organ failure [11 patients (2%)], cardiovascular [6 patients (1%)], cerebrovascular [6 patients (1%)], infections [3 patients (< 1%)], respiratory [2 patients (< 1%)], hemorrhage [1 patient (< 1%)], and other [4 patients (< 1%)].

Patients with FDCM were less likely to die compared to NICM [hazard ratio (HR): 0.720, 95% confidence interval (CI): 0.507-1.023] and ICM (HR: 0.61, 95%CI: 0.43-0.86), less likely to be delisted due to deterioration compared to NICM (HR: 0.49, 95%CI: 0.32-0.78) and ICM (HR: 0.39, 95%CI: 0.25-0.6), less likely to die or to be delisted due to deterioration compared to NICM (HR: 0.62, 95%CI: 0.47-0.81) and ICM (HR: 0.5, 95%CI: 0.38-0.66), less likely to be delisted due to improvement compared to NICM (HR: 0.28, 95%CI: 0.13-0.59) and ICM (HR: 0.35, 95%CI: 0.16-0.74), and more likely to be transplanted compared to NICM (HR: 1.25, 95%CI: 1.14-1.37) and ICM (HR: 1.83,

95%CI: 1.08-1.3) while waiting HT (Table 2).

Factors associated with waitlist mortality or delisting for FDCM on multivariate analysis patients were: mechanical ventilation (HR: 3.69, 95%CI: 1.02-13.36), creatinine (HR: 1.38, 95%CI: 1.21-1.57), and UNOS status 1A (Table 3).

Post-transplant outcomes

There was no significant difference between FDCM and other types of cardiomyopathies in stroke rates (FDCM 1.4% *vs* NICM 2.3%, *P*=0.239; *vs* ICM 3.0%; *P* = 0.051), permanent pacemaker placement (FDCM 3.6% *vs* NICM 3.4%, *P* = 0.785, *vs* ICM 3.3%, *P* = 0.681), rejection rates (FDCM 11.4% *vs* NICM 9.8%, *P* = 0.283), dialysis need (FDCM 9.7% *vs* NICM 9.5%, *P* = 0.866; *vs* ICM 10.2%, *P* = 0.806), and LOS (FDCM 17.3 ± 13.1 *vs* NICM 19 ± 22, *P* = 0.105) after HT. When compared to ICM, FDCM patients had significantly higher early rejection rates (FDCM 11.4% *vs* ICM 8.4%, *P* < 0.034), and lower LOS (FDCM 17.3 ± 13.1 *vs* ICM 20.7 ± 25.4, *P* < 0.006) (Table 4).

One, three, and five-year post-transplant survival were as follows: FDCM (91%, 88%, and 80%), NICM (91%, 84%, 79%), and ICM (89%, 82%, 75%), respectively, with no statistically significant differences between FDCM and NICM (*P* = 0.225) but higher survival compared to ICM (*P* = 0.008)(Figure 1).

DISCUSSION

Herein we describe the largest contemporary cohort of patients with end-stage heart failure from FDCM listed for HT and report on their clinical characteristics and outcomes.

Our data showed that around 2.7% of patients listed for HT have FDCM, considerably lower than the overall prevalence of FDCM. The low prevalence of the disease among patients listed for HT in our cohort might be explained by the fact that FDCM is often underdiagnosed[6].

We found that patients with FDCM who are listed for HT tended to be younger and less predominantly males compared to ICM and NICM patients, which is consistent with previous literature[7]. In addition, we found that the diagnosis of FDCM is associated with less acuity at listing, as FDCM patients were more likely to be listed as a status 2, less likely to need LVAD, and more likely to be transplanted. When FDCM patients do need MCS, they more often need biventricular support, as is illustrated by their higher usage of TAHs.

We also presented the clinical course of FDCM patients in the transplant waitlist and we showed that FDCM patients were less likely to deteriorate or die, but also less likely to improve compared to other heart failure patients. As a result, FDCM patients were more likely to be transplanted. This suggests that listed FDCM patients can be safely followed until a suitable donor is available, obviating the need for MCS as a bridge to transplant.

We also investigated transplantation outcomes in FDCM patients, which might be a concern on these patients given the fear of early rejection, as they tend to be younger with active immune system[11,12]. Previously published data compared between FDCM and non-FDCM patients who are listed for HT and showed that rejection incidence is similar in both groups[1], however, immunosuppression therapies have significantly changed since that study. We found that FDCM patients were more likely to be treated for post transplantation rejection (11.4%) compared to ICM (8.4%). That maybe explained, in part, by the fact that FDCM patients were younger and likely to have more active immune system compared to older patients[11-14].

To the best of our knowledge, our study is the largest contemporary study that compared FDCM to NICM and ICM, and followed patients after HT. We found that FDCM patients had higher survival at one, three, and five years after HT compared to ICM patients, with no significant difference compared to NICM patients. As FDCM patients were less likely to have hepatic or renal dysfunction, that may explain the higher rates of survival after HT in this group[15]. Besides that, ICM patients tend to have more comorbidities compared to patients with NICM, which may explain the higher mortality rate in ICM group[16]. Valentine *et al*. compared between FDCM and NICM and found that FDCM patients had higher survival compared to NICM patients 5 years after HT, however, the large discrepancy in sample size between the 2 groups in that study makes statistical comparison invalid[1].

Our study presents the clinical outcomes of patients with end-stage heart failure from FDCM listed for HT. The outcomes of our study may help providers in making clinical decisions while following these patients before and after HT.

Limitations of our study are mainly associated with registry-based analysis with a

Table 2 Wait-list outcomes by etiology

Outcome	FDCM *vs* NICM HR (95%CI), *P* value	FDCM *vs* ICM HR (95%CI), *P* value
Waitlist mortality	0.720 [0.507-1.023], $P = 0.067$	0.609 [0.429-0.864], $P = 0.005$
Delisting due to deterioration	0.499 [0.319-0.781], $P = 0.002$	0.387 [0.248-0.604], $P < 0.001$
Waitlist mortality or delisting due to deterioration	0.617 [0.468-0.813], $P = 0.001$	0.501 [0.381-0.659], $P = 0.001$
Delisting due to improvement	0.277 [0.131-0.588], $P = 0.001$	0.347 [0.163-0.735], $P = 0.006$
Transplant	1.248 [1.135-1.373], $P < 0.001$	1.183 [1.076-1.302], $P = 0.001$

NICM: Non-ischemic cardiomyopathy; FDCM: Familial dilated cardiomyopathy.

limited sample size of patients with FDCM, due to underestimation of the disease[6]. Therefore, our results should be interpreted in this context. Although regularly onsite audits are performed for the UNOS registry, the actual quality of the patient data has not been subject to a comprehensive audit[10]. Second, this database did not address how the diagnosis of FDCM was made and whether relatives of patients with FDCM had echocardiography to confirm the diagnosis of FDCM. Third, this registry did not mention the type of inotrope, doses, and other treatments such as: inhaled nitric oxide, or prostacyclins that were used while awaiting transplantation. Fourth, the database is missing the reason of mechanical ventilation. Although the difference of its incidence was not significant, we did not know if patients were intubated due to a cardiac etiology or any other reason. Fifth, graft failure rates might be underestimated across the groups, as its occurrence requires inotropes or mechanical ventilation support after transplantation, which is not captured by the UNOS database. Finally, as listing practices and peri-transplant care may be different in different countries, our results may not be applicable to transplant centers in other countries because UNOS is a US-based registry.

In conclusion, patients with end-stage FDCM are listed at a younger age, most often as status 2, and more frequently transplanted than patients with other cardiomyopathies. Although FDCM is associated with more frequent early rejection, survival of these patients is similar or better than other heart transplant recipients.

Table 3 Determinants of wait-list mortality or delisting

	Univariable HR (95%CI)	*P* value	Multivariable HR (95%CI)	*P* value
Age at listing		0.082		
Gender		0.413		
Ethnicity		0.712		
UNOS listing status		< 0.001		0.001
1b *vs* 1a	0.431 [0.231-0.805]		0.606 [0.305-1.204]	
2 *vs* 1a	0.160 [0.073-0.350]		0.176 [0.073-0.424]	
7 *vs* 1a	0.852 [0.252-2.886]		1.326 [0.373-4.707]	
Inotropes		0.110		
ECMO		0.229		
IABP	3.987 [1.575-10.090]	0.004		0.124
Mechanical Ventilation	4.294 [1.333-13.831]	0.015	3.694 [1.022-13.360]	0.046
VAD		0.009		0.519
LVAD *vs* no VAD	1.084 [0.540-2.179]			
BiVAD *vs* no VAD	7.636 [2.342-24.900]			
TAH *vs* no VAD	1.724 [0.236-12.600]			
ICD		0.392		
Creatinine	1.275 [1.142-1.422]	< 0.001	1.377 [1.211-1.566]	< 0.001
PASP	1.022 [1.002-1.043]	0.033		0.169
PADP		0.067		
PAMP		0.097		
PCWP		0.387		
CO		0.093		
List year		0.282		

UNOS: United network for organ sharing; ECMO: Extracoroporeal membrane oxygenation; IABP: Intraaortic balloon pump; VAD: Ventricular assist device; LVAD: Left ventricular assist device; BiVAD: Biventricular assist device; ICD: Implantable cardioverter-defibrillator; PCWP: Pulmonary capillary wedge pressure; CO: Cardiac output.

Table 4 Post-transplantation outcomes *n* (%)

	NICM	FDCM	*P*-value	ICM	*P* value
Stroke	114 (2.3)	6 (1.4)	0.239	147 (3.0)	0.051
Permanent pacemaker	171 (3.4)	16 (3.6)	0.785	164 (3.3)	0.681
Treated for rejection	496 (9.8)	51 (11.4)	0.283	416 (8.4)	0.034
Dialysis	476 (9.5)	43 (9.7)	0.866	507 (10.2)	0.806
LOS (d)	19 ± 22	17.3 ± 13.1	0.105	20.7 ± 25.4	0.006

NICM: Non-ischemic cardiomyopathy; FDCM: Familial dilated cardiomyopathy; ICM: Ischemic cardiomyopathy; LOS: Length of stay.

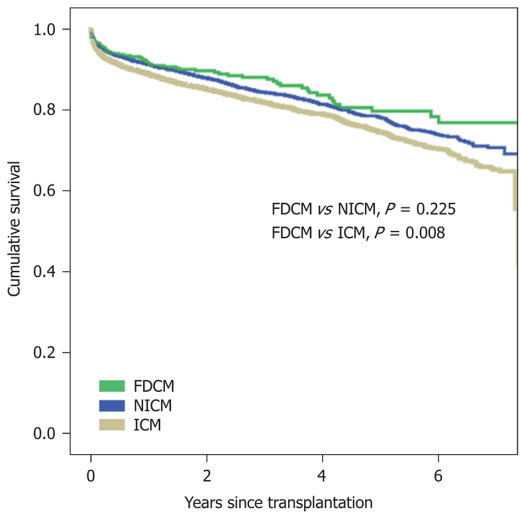

Figure 1 Kaplan-Meir graph showing post-transplant survival in familial dilated cardiomyopathy, non-ischemic cardiomyopathy, and ischemic cardiomyopathy patients. One, three, and five years post-transplant survival of patients with FDCM (91%, 88%, and 80%) was similar to those with NICM (91%, 84%, 79%, *P* = 0.225), but superior to those with ICM (89%, 82%, 75%, *P* = 0.008), respectively. NICM: Non-ischemic cardiomyopathy; FDCM: Familial dilated cardiomyopathy; ICM: Ischemic cardiomyopathy.

ARTICLE HIGHLIGHTS

Research background

Familial dilated cardiomyopathy (FDCM) is a sub-type of non-ischemic cardiomyopathy (NICM) that may lead to end-stage heart failure requiring heart transplantation (HT). This group of patients tends to develop heart failure at earlier age and they are more likely to have less comorbidity, which suggest they may have better outcomes after HT. Although characteristics of FDCM patients with end-stage heart failure have been reported, the outcomes of FDCM patients listed for HT were not described.

Research motivation

As the outcomes of FDCM listed for HT patients were not studied, we used a large database to compare FDCM to ischemic cardiomyopathy (ICM) and NICM patients who are listed for HT. Our results may help to better understand the clinical course of FDCM patients while they are awaiting HT and their outcomes after being transplanted.

Research objectives

The objective of this study was to compare FDCM to ICM and NICM patients who are listed for HT and describe their clinical course while awaiting HT and their post-HT outcomes.

Research methods

We identified patients who are listed for HT using the United Network for Organ Sharing Registry. We divided patients to three groups: ICM, NICM, and FDCM, and compared clinical outcomes of FDCM to ICM and NICM patients who are listed for HT.

Research results

FDCM patients were younger, less likely to be males, more likely to be listed as status 2, less likely to require mechanical support, but more likely to need total artificial heart. While awaiting HT, FDCM patients were less likely to die compared to ICM [HR 0.609 (0.429-0.864)], less likely to be delisted due to deterioration compared to ICM [0.387 (0.248-0.604)] and NICM [0.499 (0.319-0.781)], less likely to die or to be delisted due to deterioration compared ICM [0.501 (0.381-0.659)] and NICM [0.617 (0.468-0.813)], less likely to be delisted due to improvement compared to ICM [0.347 (0.163-0.735)] and NICM [0.277 (0.131-0.588)], and more likely to be transplanted compared to ICM [1.183 (1.076-1.302)] and NICM [1.248 (1.135-1.373)]. After HT, FDCM patients were more likely to have early rejection compared to ICM (FDCM 11.4% *vs* ICM 8.4%; $P < 0.034$), but more likely to survive (91%, 88%, and 80%) compared to ICM (89%, 82%, and 75%) at 1, 3, and 5 years, respectively.

Research conclusions

Patients with end-stage heart failure due to FDCM are more likely to be transplanted compared to NICM and ICM. After HT, they are more likely to develop early rejection, but more likely to survive compared to ICM patients.

Research perspectives

This study may help providers in making clinical decisions for patients with end-stage heart failure due to FDCM while waiting and after HT.

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