Clinical profile and outcomes of childhood dilated cardiomyopathy – A single-center three-decade experience

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ABSTRACT

Introduction and Aims : **Dilated cardiomyopathy (DCM) is an important cause of heart failure (HF) among children. Research on pediatric DCM remains surprisingly scarce. The primary objective of the study was to evaluate the clinical profile and outcomes of pediatric DCM and the secondary objective was to study the predictors of outcome.**

Methods and Results : **We enrolled all patients with cardiomyopathy who presented to us between 1990 and 2020 and were younger than 18 years. During the 30-year study period, we identified 233 cases of pediatric cardiomyopathy. One hundred and nineteen (51%) cases had DCM. This retrospective cohort was analyzed to study their outcome and the possible predictors of outcome. Nearly, 8% presented in the neonatal period, and 37% in infancy. The most common mode of presentation was dyspnea on exertion (71%). Ninety-three patients presented in heart failure (78%). The median left ventricular dimension z-score in diastole was 4.3 (range 2.5–9.06). The median left ventricle (LV) ejection fraction was 31%. Seventy-two percent of this cohort were on angiotensin-converting-enzyme inhibitors, 40% on aldosterone antagonists, and 47% on beta-blockers. One-third had syndromic, metabolic, genetic, or any secondary cause identified. Twenty-seven patients satisfied the three-tiered clinical classification for the diagnosis of probable acute myocarditis. Over a mean follow-up of 3.29 years, 27% were lost to follow-up. Among the remaining patients who were on follow-up (n = 86), 39 (45%) died, 31 (36%) recovered, and 16 (18%) had persistent LV dysfunction. Heart Failure was the most common cause of death. Eight patients in this cohort (4.2%) had thromboembolic phenomena. Nine had sustained ventricular arrhythmias and six had atrial/junctional arrhythmias. Among the various risk factors studied, only infantile onset had a significant relationship with death or ventricular arrhythmias (P value– 0.05). The 5-year survival rate of DCM patients was 59%.**

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Conclusion : **A reasonably good percentage of our population showed recovery of the left ventricular function (36%). Only infantile onset had a significant relationship with death or ventricular arrhythmias. The outcome in our DCM cohort is similar to other population cohorts.**

Keywords : Dilated cardiomyopathy, India, outcomes, pediatric

INTRODUCTION

Dilated cardiomyopathy (DCM) remains the most common form of cardiomyopathy and the reason for cardiac transplantation among children. Pediatric cardiomyopathies often occur in the absence of comorbidities; and thus, offer insights into the pathogenesis of myocardial dysfunction. A better understanding of the spectrum and outcomes of childhood cardiomyopathy would facilitate patient care and permit the evaluation of newer therapies. Previous studies have described the clinical profile, outcomes, recovery of the left ventricular function, and the risk factors of sudden cardiac death (SCD).[1-4] Research on pediatric DCM as compared to adult DCM, especially from this part of the world is still limited and that was the motivation behind the conduct of this study.

MATERIALS AND METHODS

The study enrolled all patients with cardiomyopathy who presented to us between 1990 and 2020 and were younger than 18 years old. After taking informed consent, we reviewed the available medical records of each enrolled patient. This retrospective cohort of children was analyzed to study their outcome and the possible predictors of outcome. The primary objective of the study was to evaluate the clinical profile and outcomes of pediatric DCM and the secondary objective was to study the predictors of outcome. Exclusion criteria were prior exposure to anthracyclines or other cardiotoxic drugs, cardiomyopathy related to chronic arrhythmia, primary renal disorder, kawasaki disease, congenital heart defects not associated with malformation syndromes, any history of rheumatic fever, HIV Infection, and cardiothoracic or any other major surgery. After directly reviewing all available cardiac information, we assigned each patient to a diagnostic category according to the phenotypic characteristics following the European Society of Cardiology Classification.[5] Cardiomyopathies were defined as abnormalities of the ventricular myocardium unexplained by abnormal loading conditions or congenital heart disease.[6] DCM was defined as LV end-diastolic dimension >2 standard deviations above normal for body surface area, in conjunction with depressed systolic function.[7] Normalization of LV function was defined as a return to normal LV size (an LVEDD z-score <2) and normal LV systolic function (an left ventricular fractional shortening (LVFS) or LV ejection fraction more than 55%) on any subsequent echocardiogram.[8] Since there are no specific recommendations for implantable cardioverter-defibrillator (ICD) implantation in the pediatric population according to guidelines, adult criteria were applied. ICD was implanted in the presence of hemodynamically not tolerated VT or VF as a class Ia Indication.[9] Need for cardiac resynchronization therapy (CRT) was defined as those who meet current recommendations for the implantation of a CRT as in adults with the New York Heart Association class II to ambulatory class IV heart failure (HF), left ventricle ejection fraction (LVEF) <35%, and widened QRS.[10] Pulmonary artery hypertension on echocardiography was defined by elevated mean pulmonary artery pressures of more than 20 mmHg as assessed by the peak pulmonary regurgitation jet. The left ventricular ejection fraction and LV end-diastolic dimension were calculated by M mode echocardiography for all patients since LVEF by the Simpsons was not available for all. Collected data include demographic descriptors, relevant history, examination findings, and laboratory data. Patients were considered on follow-up if they were seen in the outpatient department in the previous 6 months. If not seen within the past 6 months, patients or their guardians were contacted by telephonic interview. A single observer read the earliest available electrocardiography (ECG) and converted the measurements to age-appropriate Z-scores.[11]

Echocardiographic measurements of the left ventricular dimensions, diastolic-free wall, and septal thickness were expressed as Z-scores based on the body surface area.[12] The outcome parameters to be studied were age at death, circumstances of death (Sudden cardiac death, death from ventricular arrhythmia, or death from HF), frequency of atrial, junctional, or ventricular arrhythmias, atrioventricular block, Bundle branch block, and requirement of ICD/CRT/transplant. The study was approved by the institutional ethical committee of the center. For patients, not on follow-up, the need for informed consent was waived by the hospital ethics committee.

Statistical analysis

The data analysis was performed using the SPSS Statistics software for Windows Version 21. Continuous variables were expressed as either mean ± standard deviation, or median, depending on the overall variable distribution.

Descriptive summaries were presented as frequencies and percentages for categorical data. Continuous variables were compared using Student's *t*-test or Mann–Whitney U test as appropriate.

RESULTS

The clinical profile of DCM patients is presented in Table 1. Ten of the 119 DCM patients (8%) presented in the neonatal period, and 45 (37%) presented in infancy. The most common mode of presentation was dyspnea on exertion (71%). Ninety-three patients presented with HF (78%). ECG characteristics of the DCM patients are described in Table 2. Out of the four patients with preexcitation, only one of them had a documented orthodromic atrioventricular reentrant tachycardia (AVRT), the rest had no documented arrhythmia. The pathway was localized to the right anteroseptal region in one, the mid-septal region in another, and to the right atrioventricular groove in two, based on the 12-lead ECG. Echocardiographic characteristics are described in Table 3. None of the patients had regional wall motion abnormalities as assessed on echocardiography. The right ventricle dysfunction in addition to LV dysfunction was noted in 10 patients. Cardiac magnetic resonance imaging could be done only for 24 patients as it would mostly require general anesthesia and a small number of patients consented to it. Among them, late gadolinium enhancement (LGE) was present in 10 (41%). Three had subendocardial, three transmural, two midmyocardial, one had patchy, and one had a mixed type of enhancement. One patient had LGE of >50%. LGE was found in the septal wall of basal or midsegments in five patients, the lateral wall in three and the anterior wall in two. Two patients satisfied the revised lake louis criteria for the diagnosis of myocarditis.[13]

Holter monitoring was done only for 11 patients. Four had nonsustained ventricular tachycardia/ventricular premature complexes, one had atrial tachycardia, one had BBB, and the rest had a normal scan. 74 % of the DCM patients were on digoxin, 89% were on furosemide, and 72% were on angiotensin-converting-enzyme inhibitors (ACE-I) at admission or anytime during follow-up. Only 40% were on aldosterone antagonists, and 47% were on beta-blockers. Digoxin was prescribed in the early period of the study, and the practice of prescribing digoxin fell in the later part. ACE-I and beta-blockers were more commonly prescribed in the latter half of the study period. Only six patients were on amiodarone, and seven were on warfarin. Six were started on warfarin for treatment of the thromboembolic phenomenon and one was started prophylactically given severe LV dysfunction. Two patients were started prophylactically on aspirin and one following a cerebral embolic event. The practice of prescribing ARNI started recently; thus, only four were on ARNI and two were on ivabradine. Five patients received intravenous immunoglobulins for myocarditis. Sixteen patients were catheterized in the early periods of the study at the time

Table 1: Clinical characteristics of dilated cardiomyopathy patients

Variable (*n*=119)	Frequency (%)
Age at diagnosis	
<1 month	10 (8)
1-12 months	35 (29)
1-12 years	48 (40)
>12 years	26 (21)
Male/female	69/50 (57)
Mode of presentation	
DOE	84 (71)
Febrile illness	13 (11)
Screening	7 (5)
Recurrent RTI	8 (7)
Incidentally	4 (3)
Arrhythmias	2 (1.6)
Syncope	3 (2.5)
Family history of cardiomyopathy	14 (11)
Family history of SCD	11 (9)
Consanguinity (*n*=106)	18
Syndromic	7 (5) (1 DMD, 1 BMD, 1 KS, 1 MPS Type 6, 2 MD, 1 US)
Metabolic	6 (2 primary carnitine deficiency, 4 severe Vitamin D3 deficiency)
Probable acute myocarditis	27
FC	
I	6 (5)
II	64 (53)
III-IV	44 (41)
Cardiac enlargement	90 (75)
LVS3	64 (54)
Loud P2	48 (40)
ESM/PSM	61 (51)
CXR	
CTR, mean±SD	0.65±0.06
LAE	79 (73)
PVH	94 (87)

DOE: Dyspnea on exertion, RTI: Respiratory tract infection, SCD: Sudden cardiac death, CXR: Chest X-ray, CTR: Cardiothoracic ratio, ESM/PSM: Ejection systolic murmur or pan systolic murmur, PVH: Pulmonary venous hypertension, FC: Functional class, LAE: Left atrial enlargement, P2: Pulmonary component of second heart sound, KS: Klinefelter syndrome, US: Unclassified syndromes, MD: Mitochondrial disorder, DMD; Duchenne muscular dystrophy, BMD: Becker muscular dystrophy, MPS: Mucopolysaccharidosis type 6, SD: Standard deviation

Table 2: Electrocardiogram characteristics of dilated cardiomyopathy patients

ECG (*n*=119)	Median (range)/*n* (%)
PR interval (ms)	140 (80-240)
PR interval (Z score)	1.0 (–2.0-4.7)
QRS d (ms)	100 (80-180)
QRS d (Z score)	2.0 (–1.0-7.3)
LAD	13 (12)
RAD	4 (3)
Normal axis	102
Q wave	41 (34)
ST depression	2 (1.6)
LVH	96 (80)
Preexcitation	4

LAD: Left axis deviation, RAD: Right axis deviation, LVH: Left ventricular hypertrophy, ECG: Electrocardiogram

of diagnosis. The mean PVRI was 4.3 ± 3.9 WU.m^2, PA mean was 26 ± 11 mmHg, and cardiac index was 2.7 ± 1.4 L/min/m^2. Forty (33%) patients had syndromic, metabolic, or any secondary cause identified [Table 1]. Viral identification was available only for three patients. One had herpes zoster viral myocarditis, and two had coxsackie viral analysis positive. None of this cohort underwent endomyocardial biopsy. Vitamin D3 levels were checked in 12 patients among whom four had levels <30 ng/ml and four had levels <20 ng/ml. All eight patients were started on Vitamin D supplementation, of whom three recovered. Serum thyroid-stimulating hormone was checked in 42% of patients, of whom only two had hypothyroidism and were given thyroxine replacement. One patient recovered after supplementation. Table 4 summarizes the follow up echocardiographic data of the subjects who died, those who had persistent LV dysfunction and those who improved.

Outcome

The outcome characteristics are described in Table 5. Over a mean follow-up of 3.29 years ± 4.73 (range 0.1–24 years), 27% were lost to follow-up. Among the remaining patients who were on follow-up (n = 86), 39 (45%) died, 31 (36%) recovered, and 16 (18%) had persistent LV dysfunction. Eight patients in this cohort (6.7%) had thromboembolic phenomena either at presentation or during follow-up. Two patients had left ventricular thrombus in the absence of embolic events, three patients had a superficial femoral artery embolism, and three had cerebral emboli. One of the patients with LV thrombus had an associated atrial flutter.

Among the 119 DCM patients, nine had sustained ventricular arrhythmias either at presentation or during follow-up. Among the patients with sustained ventricular arrhythmias, six died, one was alive and received an ICD, and two were lost to follow-up. Atrial/junctional arrhythmias were present in six patients. One patient each had atrial fibrillation, atrial flutter, and paroxysmal atrial tachycardia on follow-up. One patient had an orthodromic AVRT secondary to a parahisian accessory pathway on follow-up. One patient had supraventricular tachycardia, and one had junctional tachycardia.

The 5-year survival rate of DCM patients was 59% [Figure 1]. Among the various risk factors studied, only infantile onset had a significant relationship with death or ventricular arrhythmias (P = 0.05) [Table 6]. Table 7 compares the patients with a history of probable acute myocarditis with others.

Table 3: Echocardiographic characteristics of dilated cardiomyopathy patients

Echocardiographic characters (*n*=119)	Median (range)/*n* (%)
LVId d (mm)	48 (30-84)
LVId d (Z score) (*n*=98)	4.3 (2.5-9.06)
LVId s (mm)	40 (25-71)
LVId s (Z score) (*n*=98)	5.8 (2.6-10.2)
LVEF (%)	31 (10-47)
Moderate or severe PAH	28 (23)
Mitral regurgitation (*n*=104)	
Mild	46 (38)
Moderate	53 (44)
Severe	5 (4)

PAH: Pulmonary artery hypertension, LVEF: Left ventricular ejection fraction, LVId d: Left ventricular internal dimension in diastole, LVIds: Left ventricular internal dimension in systole

Table 4: Depicts the echocardiographic follow-up data of the three groups of patients

	Subjects who died (*n*=39)		Subjects with persistent LV dysfunction (*n*=16)		Subjects who improved (*n*=31)	
	Baseline	Follow up	Baseline	Follow up	Baseline	Follow up
Mean EF (%)	20	22	38	33	34	52

EF: Ejection fraction, LV: Left ventricle

Table 5: Outcome in dilated cardiomyopathy patients

Outcome	*n* (%)
Years of follow-up, mean (range)	3.29±4.73 (0.1-24)
Lost to follow-up (*n*=119)	33 (27)
Death (percentage of patients on follow-up, *n*=86)	39 (45)
Death from HF	28
SCD	6
Death from ventricular arrhythmias	5
Alive (percentage of patients on follow-up, *n*=86)	47 (54)
Recovered (%)	31 (36)
Persistent LV dysfunction (%)	16 (18)
Sustained ventricular arrhythmias	9 (7.7)
Atrial/junctional arrhythmias	6 (5.1)
AV block	14 (11.9)
BBB	9 (7.7)
LBBB	2
Requirement of ICD/CRT/transplant	41 (34)
ICD/PPI/CRT	1 ICD, 2 CRT
ECMO	1
EPS	1
Thromboembolic phenomena	8 (6.7)

DCM: Dilated cardiomyopathy, HF: Heart failure, SCD: Sudden cardiac death, LV: Left ventricular, BBB: Bundle branch block, LBBB: Left BBB, ICD: Implantable cardioverter defibrillator, CRT: Cardiac resynchronization therapy, PPI: Permanent pacemaker implantation, ECMO: Extracorporeal membrane oxygenation, EPS: Electrophysiology studies, AV: Atrioventricular

DISCUSSION

This is the most extensive study on pediatric cardiomyopathy from South Asia as of now. About 37% of this DCM cohort presented in infancy, and 33% had syndromic, metabolic, genetic, or any secondary cause identified. The 5-year survival rate was 59%. Thirty-six percent showed recovery of the left ventricular function. Among the various risk factors studied, only infantile onset had a significant relationship with death or ventricular arrhythmias.

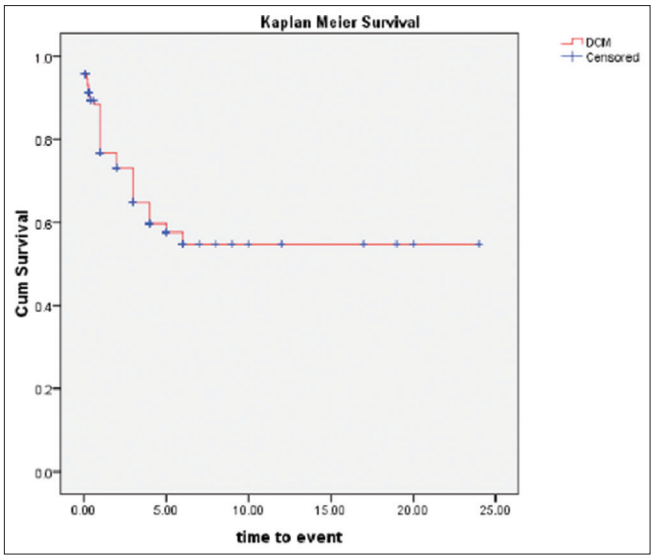

Figure 1: Kaplan–Meir curve showing survival in the DCM patients. The Kaplan-Meir survival curve reveals 5-year survival rate of 59% in DCM patients. DCM: Dilated cardiomyopathy

Table 6: Depicts the relationship between various variables and death and/or ventricular arrhythmias

Variable	*P*
LVId Z score >4	0.63
Age, infancy versus noninfancy	0.05
Family history of cardiomyopathy	0.76
Syncope	0.60

LVId: Left ventricular internal dimension

Table 7: Compares the subjects with a history of probable acute myocarditis with others/patients who did not have such a history

	Postmyocarditis (*n*=27), *n* (%)	No history of myocarditis (*n*=92), *n* (%)	*P*
Age at diagnosis (mean)	5.5	5.6	0.51
Infantile onset	8 (29)	35 (38)	0.63
Consanguinity	5 (18)	13 (14)	0.71
Family history of cardiomyopathy	1 (3)	13 (14)	0.20
LVId Z-score >4	12 (44)	45 (48)	0.80
Years of follow-up (mean)	3.4	3.2	0.71
Mortality	8 (29)	31 (33)	0.62
SCD	2 (7)	4 (4)	0.58
Recovered	11 (40)	20 (21)	0.10

LVId: Left ventricular internal dimension, SCD: Sudden cardiac death

Researchers from other parts of the world have studied the outcomes and clinical features of pediatric DCM in detail. Alexander *et al.* examined the long-term outcomes for 175 children with DCM <10 years enrolled in the NACCS registry.[1] About 64% of the patients presented in infancy in his cohort as compared to 37% in our cohort. The comparatively lesser incidence of infantile DCM may be related to less frequent screening for inborn errors of metabolism (IEM) and familial DCM in our cohort. About 8.9% of his cohort had a metabolic or mitochondrial disease identified. In our study, 13 patients had proven syndromic or metabolic disease [Table 1]. Syndromic or metabolic disease must have been underestimated as all did not undergo screening for the same. In this cohort, one patient had Duchenne muscular dystrophy and one had Beckers muscular dystrophy, both confirmed by genetic testing. One patient with a positive family history of DCM in another sibling presented in the neonatal period with feeding difficulty. She was evaluated and found to have carnitine deficiency. Both siblings responded to carnitine supplementation. Diagnosing such disorders, especially carnitine deficiency, is essential as appropriate treatment reverses cardiomyopathy. Vitamin D3 levels were checked in 12 patients among whom four had levels <30 ng/ml and four had levels <20 ng/ml. In the study by Raafat and EL-Asheer. (n = 44), Vitamin D deficiency was found in 90.9% of children with idiopathic DCM.[14] We acknowledge that fewer patients had Vitamin D3 levels checked which may be due to financial constraints. Among 119 DCM patients, 27 satisfied the three-tiered clinical classification for the diagnosis of probable acute myocarditis,[15] but none of the patients underwent endomyocardial biopsy to diagnose definite myocarditis. In the study by Daubeney *et al.*, a potential viral contribution was identified in 68.2% of 44 cases who underwent cardiac histological examination.[2]

Forty-seven percent of patients were on beta-blockers, and 72% were on ACE-I, showing that a reasonably good percentage of our population was on beta-blockers. Only 5% of the pediatric patients enrolled in the American Pediatric Cardiomyopathy Registry received beta blockers in the 1990s compared with 18% after 2000.[16] Beta-blocker use in the pediatric population has been increasing in recent times. Although Shaddy *et al.* in their study observed that carvedilol does not significantly improve clinical HF outcomes in children and adolescents with symptomatic systolic HF, the study may have been underpowered and the effect may have been due to the inclusion of right ventricle and single ventricle physiology patients.[17] Eight patients in this cohort (6.7%) had thromboembolic phenomena. Two had left ventricular thrombus in the absence of embolic events, three had a superficial femoral artery embolism, and three had cerebral emboli. All except two had severe LV dysfunction with LVEF <20%. None of the subjects with thromboembolic phenomena were on prophylactic anticoagulation. According to the review published by Chen *et al.*, The incidence of intracardiac thrombosis in pediatric DCM ranges from 6% to 53% and that of embolism ranges from 1% to 16%. The left ventricular ejection fraction below 25% is a major risk factor for intracardiac thrombus formation in the pediatric DCM population.[18] In the largest pediatric study by Gunthard *et al.* (n = 130), 9% of patients without prophylactic

anticoagulation (during follow-up or before the first embolic event) experienced an embolism and among all, 14% showed evidence of thrombosis or embolism.[19] Only two of our patients were on ivabradine and four were on ARNI. In recent studies, ivabradine was shown to reduce the resting heart rate of children with chronic HF and DCM and improved left ventricular ejection fraction, clinical status, and quality of life.[20] ARNI has already been approved by FDA for pediatric HF after the encouraging early results of the PANAROMA-HF study.[21] The fewer number of patients on such novel therapies in our study highlights the need for rapid adoption of such practices in our population.

Among the DCM subgroup, over a mean follow-up of 3.29 years, 33 were lost to follow-up (27%). Among the remaining 86 patients, 39 (45%) died, and 47 (54.6%) were alive. Among the alive patients, 31 (36%) recovered, and 16 (19%) had persistent LV dysfunction. Patients who recovered were in functional class I-II on follow-up and children who had persistent LV dysfunction were in the same functional class as they were at the time of diagnosis. Percentage of patients showing recovery of LV function is almost similar to other studies conducted so far. In the study by Alexander *et al.*, 33% of all patients attained normalization of LV function 15 years after diagnosis.[1] Higher LVFS Z scores at diagnosis, higher LVFS Z scores during follow-up, and more significant improvement in LVFS Z scores were all predictive of an increased likelihood of normalization. Arola *et al.*, in their study, observed that during a mean follow-up of 3.9 ± 4.5 years (range, 1 day to 25 years), 10 patients (16%) recovered, 17 (27%) had a residual disease, 4 (6.4%) underwent heart transplantation, and 31 (50%) died.[3] Patients who had normalized LV function most likely would have viral myocarditis or have undergone reverse remodeling as an effect of medications.

The 5-year survival rate of 59% is almost similar to that of most other studies. Alexander *et al.* reported freedom from death or transplantation to be 65% at 5 years among the NACCS participants.[1] In the PCMR registry, freedom from death at 5 years in the DCM cohort was 78%, and freedom from death or transplant was 57%.[22]

BBB was present in nine (7.7%) patients, and among them, left bundle branch block (LBBB) was present in only two patients. This brings attention to the lower incidence of LBBB in pediatric DCM compared to adult DCM as has been proven in previously conducted studies. The study by Puggia *et al.* reported on the natural history of DCM in children. Out of the 927 DCM patients, 47 were pediatric DCM (5.1%). LBBB was seen in 4.4% of pediatric DCM versus 31.9% of adult DCM.[23] This has been postulated to be due to the less severe disease at baseline and earlier diagnosis due to screening.

Among the various risk factors studied, only infantile onset had a significant relationship with death or ventricular arrhythmias. This may reflect IEM with infantile onset as the underlying etiology of DCM in such cases which portends a worse prognosis. In the study by Alexander *et al.*, age at diagnosis <4 weeks or >5 years, familial cardiomyopathy, and lower baseline LVFS z score were associated with increased risk of death or transplantation.[1] About 34% of patients fulfilled the criteria of requiring advanced HF therapies such as CRT/ICD or cardiac transplant. However, only three patients underwent CRT or ICD and none underwent a transplant. We must acknowledge that access to these therapies is still limited in South Asia because of financial reasons. Table 7 compares the patients with a history of probable acute myocarditis with others. As expected, infantile onset and family history of cardiomyopathy are less common in the myocarditis group. This reflects the genetic and metabolic causes which have an infantile onset, being less common in this cohort. The myocarditis group has lesser mortality and a greater number of patients who have recovered.

The strength of the study is that this is the largest study conducted till now from south Asia on pediatric cardiomyopathies. The limitation is its descriptive, retrospective nature. A significant number of patients have been lost to follow-up. That is expected in a retrospective study conducted over three decades. Fewer patients were screened for metabolic and genetic disorders, due to financial constraints, which make the cohort heterogeneous.

CONCLUSIONS

The 5-year survival in our DCM cohort is almost similar to other populations (59%). Among the patients on follow-up (n = 86), 45% died, and 18% had persistent LV dysfunction. A reasonably good percentage of our population showed recovery of the left ventricular function (36%). About 7.7% had sustained ventricular arrhythmia and 5.1% had atrial or junctional arrhythmias. About 22% had a history of probable acute myocarditis. Eight patients had Vitamin D3 deficiency and three of them recovered after supplementation. Only infantile onset had a significant relationship with death or ventricular arrhythmias.

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Conflicts of interest

There are no conflicts of interest.

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