Clinical profile of childhood dilated cardiomyopathy in Tripoli children hospital

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Abstract: Dilated cardiomyopathy (DCM) in children is a progressive disease with high morbidity and mortality; we studied the clinical presentation and their outcome with dilated cardiomyopathy (DCM). **Methods:** retrospective study of medical records of patients diagnosed and follows up with dilated cardiomyopathy in cardiac clinic Tripoli children hospital from December 2006 till December 2010, follow up period from one day to four years. They were 45 patients (one patient excluded CHD) underwent clinical examination, chest X ray, ECG, and echocardiograph. Sex, age at presentation and mode of therapy all were analyzed. **Results:** total number of patients was 45 patients one patient excluded. Male to female ratio was 1.5 : 1, 60% males, 40% females, 32 patients ≤ 12 months 73 % (of these 75%, 24 cases were less than 6 months). The majority of cases presented with heart failure, eleven patients died 25% (73%, 8 cases, diseased during the first 12 months). Males had poor prognosis. **Conclusion:** the risk age group was first 12 months of age, males had poor prognosis, and this disease needs more study to explore the causes and early management to avoid high mortality in this age groups.

keywords: dilated cardiomyopathy, echocardiograph, Libya.

Introduction

Dilated cardiomyopathy is not uncommon in pediatric age group with high morbidity and mortality in children, dilatation of the heart leading to impaired contractility and supervene heart failure. DCM caused by varies agents, idiopathic is the most common one, viral infections play an important role in getting DCM, nutritional deficiencies, severe anemia, metabolic, toxins and endocrine disorders like hyperthyroidism or hypothyroidism. The World Health Organization (WHO) in 1995 classified cardiomyopathies into dilated, hypertrophic, restrictive, arrhythmogenic right ventricular and unclassified types (1).

The commonest type of cardiomyopathy is DCM (58%), hypertrophic type (30%), restrictive type (5%) and arrhythmogenic right ventricle (ARVC) (5%) (2). DCM defined as cardiac dilatation mainly left side especially left ventricle and decreased systolic function. DCM is characterized by dilatation of left ventricle with impaired contractility. The PCMR, the annual incidence of DCM in children < 18 years old was 57/100 000 per year, was higher in boys than in girls (0.66 versus 0.47/100 000), in blacks than in whites (0.98 versus 0.46/100 000) and in infants than in children (4.40 versus 0.34 /100 000). (3)

Dilated cardiomyopathy is a disease of myocardium causing dilatation of the left side of the heart mainly left

ventricle with normal septal and left ventricle free wall thickness, (in some cases can affect both ventricles) which leading to impairment of its systolic function and subsequently heart failure occurs. The diagnosis based on clinical presentation of heart failure and impaired left ventricle function which is confirmed by echocardiography finding with low shortening fraction and low ejection fraction with poor contractility and dilatation of the left ventricle with or without mitral regurgitation.

Materials and methods

This is a retrospective study between December 2006 and December 2010. All patients were diagnosed with DCM and follow up in cardiac clinic Tripoli children hospital for four years ranging from intrauterine (during fetal echocardiography) to four years. Age at presentation ranged from 5 days to 7 years. All cases underwent clinical examinations chest X ray (cardiomegaly and pulmonary congestion), ECG (ST Changes and signs of left ventricle hypertrophy), and echocardiography M mode to measure left ventricle end diastolic diameter, left ventricle end systolic diameter, SF and EF, Doppler and color Doppler to detect any valve regurgitation (we were use Phillips Envisor machine), poor contractility, left ventricle dilatation with reduced systolic function.
Patients with congenital heart diseases (e.g. anomalies coronary artery, patent ductus arteriosus) and those with a history of drugs toxicity or metabolic disorders were excluded. Follow up interval after diagnosis (0 time at diagnosis), were at one month, 6 months, one year, two years, three years and four years.

Therapy given; at presentation: in the form of a combination of intravenous immunoglobulins (IV IG), frusamide, Angiotensin converting enzymes inhibitors (ACE), and/or digoxin. Long term treatment includes diuretics, ACE, and/or digoxin. Twenty-one patients given IV IG in addition to the classical treatment of heart failure (as mentioned earlier).

Data collection: the data was collected from medical files in cardiology clinic; our patients were referred from different sources (our emergency department, in patients department, private clinic and rural hospital) for diagnosis and follow up. Age, gender and clinical presentations, therapy given and outcome all were analyzed.
Data analysis: incidence, mortality rates, data input to Microsoft excel.

Results

During the 4 years study: Patient's number and Gender: Total number of patients was 45 patients one patient excluded. M : F ratio was 1.5 : 1 (60% (27) males and 40% (18) females), figure 1. Twenty four patients were < 6 months (55%). 32 patients < 12 months (73%). Twenty-one patients (48%) presented with cardiac symptoms (tachycardia, cyanosis).

Table 1: Distribution of children with cardiomyopathy

Out come								Acute treatment				Gender		Age in months	No
Missed		Died		Still on treatment		Normal		Digoxin	ACE	Frusamide	IVIG 21 pt	F	M		
F	M	F	M	F	M	F	M				M F				
1	1	3	5	2	5	2	5	+	+	+	5 7	8	15	0 6	23
1	0	1	1	1	2	1	1	+	+	+	2 4	4	4	>6 12	8
0	3	0	1	1	0	4	3	+	+	+	1 2	5	7	>12	12

three patients (7%) of them presented with other symptoms (failure to gain weight and fever) as shown in table 1.
Male: female, from this table the males were predominantly presented with cardiac symptoms especially with younger age, sixteen male patients presented in the first six months of age (36%) and female accounts only (18%). This will show that DCM more common in males, respiratory symptoms was same in both sexes. 29 patients were presented between August and January (64.4%) then the most patients presented in autom and winter which is the session of viral infection, this may explain the improvement of the patients who were received IVIG, could be inflammatory DCM post viral myocarditis, in spite of No myocardial biopsy for detection of virus. From December 2006 to December 2007 were 18 cases, 15 cases in 2008, 9 cases in 2009 and only 2 cases in 2010. Then the peak rate was on first year of study and the least on the last year.

Acute and long term treatment: in acute stage twenty one patients received IVIG, frusamide, ACE, with or without digoxin, other twenty three were received frusamide, ACE with or without digoxin. From this therapy, we found sixteen patients (36%) return to normal heart function (10 patients received IVIG), 10 patients (23%) stationary and still on treatment (5 patients received IVIG), 11 patients died (25%) (only one received IVIG) and unfortunately seven patients (16%) missed follow up (5 patients received IVIG) our patients who were improved with IVIG it could be inflammatory DCM (post viral myocarditis).
Long term therapy was frusamide, ACE with or without digoxin, one patient has atrial flutter and put on amedarone

Mortality rate: mortality rate was 25%, three patients died after first visit all were males (6.8%) none did receive IV IG, Four patients died after one month from first visit (9.1%) ALL were males. Only one patient received

IG. Two patients died at 6 months of follow up (4.5 %) one females and one male. One female died at 2 years follow-up (received IG) (2.3 %). One died at 3 years of follow up did not receive IG (2.3%). Nine patients died with presenting age < one year (82% of total death). 21 patients received IG (45.6%). Only two patients received IV IG (died).

Drop rate: A total of seven patients missed follow up during the whole study period (15.9 %). During the first year six patients, five males and one female missed follow up (drop rate 13.6 %.). During the second year only one female missed follow up (drop rate 2.3 %). During the third year nil missed follow up (drop rate 0%). During the fourth year nil missed follow up (drop rate 0%).

Echocardiography findings: Sixteen patients showed complete recovery of their ventricular function (good ventricular contractility, shortening fraction (SF) > 28%, ejection fraction (EF) > 60% and no dilatation of the left ventricle) after 2 years of follow up (36.4%). Ten patients (22.7%) still on treatment (frusamide, digoxin and ACE) because clinically unwell, poor weight gains, tachyapnic and distressed with echocardiography finding; poor contractility, dilated left ventricle, low ejection fraction (EF) and low shortening fraction (SF). In patients who presented with very low SF or EF did not play rule in good or poor prognosis, our normal 16 patients their SF ranging from 7% to 23% and EF varies from 21% to 40%, and in died patients SF was 10% to 25% and EF varies from 22% to 51%, that means low EF or SF at presentation were not predict in the prognosis of DCM.

From above table, we found that most of the patients presented in the first 6 months of age (52.3%), eight patients died 35%, (not received IVIG) 5 males and 3 female, five male and 2 females patients recovery completely their ventricular function, seven patients still on treatment and two patients missed follow up. Other peak after 12 months of age seven males and five females, seven of them become normal (58.3%), only one male died and three missed follow up. The mortality rate was (25%). Death after first visit 6.8% which constitutes 27% of the total deaths.

The males had poor prognosis with death rate were 64%, only one patient received IVIG. Most of them were under 12 months of age. One of them with positive family history younger age groups (< 6 months) has worse prognosis mortality rate from total death was 82%. The male gender and younger age are predictor factor for poor outcome. We noticed that the peak age presentation was in the first 6 months of age, and the least was between 12-18 months of age. One quarter died (25%), more than one third return back to normal (36%) and (23%) still on treatment but unfortunately 16% missed .mortality was high in the first 12 months of age.

Discussion

Dilated cardiomyopathy is the comm.-onset acquired heart disease in child-ren. DCM is the most common type of cardiomyopathies, it accounts 55% to 58%, from cardiomyopathies, and it has high mortality and morbidity rates (2, 3). DCM can be caused by infections, metabolic disorders like hyperthyroidism, hypothyroidism,

toxins, carnitine deficiency, nutritional defic-iency, and drugs. Could be genetic or familial, Idiopathic DCM which carries a poor prognosis, but the commonest cause is post viral myocarditis, it accounts 27% of dilated cardiom-yopathy (4, 5). The current study is not determine the cause of dilated cardiomyopathy and we study all patients presented with symptoms and signs of heart failure with cardio-megaly, echocardiography revealed dilated left side of the heart with poor contractility with low shortening fraction and ejection fraction (it was < 25% and 40%) respectively .

Dilated cardiomyopathy post myocar-ditis has better prognosis than other causes of dilated cardiomyopathies (6) this could explain the good prognosis for those patients received IVIG. Males were predominance in presenta-tion (60%) which was same as in Egypt and Kuwait (7), and same in Indian children which revealed male were dominant, with same mortality rate (25%) and the risk age younger than 6 months. In other study female gender were high risk with death age < 1 year (6, 8), other study with male was also dominant and opposite in Saudi Arabia females accounts (63.9%) and males (39%) both in Jeddah and in Riyadh, 73% of presentations were before 12 months of age of our children, in comparison in Saudi Arabia 50% presented before 14 months of age, older the age worse the outcome (9). High incidence of DCM in children below one year and males > females were found in Towbin study (5). In Saudi Arabia study outcome 48.2% improved, 27.7% stationary and 24.1% deteriorated (9). In our study, 36% improved, 23 % stationary and 25% died, their patients improvement better than our patients with nearly same results in stationary patients and the same in mortality rate, therefore, in other study, the improved patients (37%) same as our patients, stationary patients (55.5%) more than us, with very low death rate (7.4%) and in Bostan, 52.5% recovery more than half of the patients, 42.5% stationary and 5% died, good prognosis is related to age ≤ 2 years (9, 10).

In other studies, it has been found that after a 2.5 years of median follow-up period, about one-third of patients fully improved near our patients, 38% survived and had left ventricular dysfunction and 29.4% died, mostly in the first year of follow-up (11). In our study 36% improved and back to normal heart, 23% stationary still on treatment which was less than Jarallah study, 25 % died which was more than his study. Other study shows good prognosis is related to the age at presentation ≤ 2 years and the patients with intractable heart failure has poor outcome which is not same as current study were found that the younger the patient the grave the prognosis and comparable with other study that the most dangerous period was the first 2 years of age (11, 12). Few study were given IVIG to the patients with recent onset of DCM (inflammatory cardio-myopathy) and shows improvement in Lf ventricle EF, IVIG specific treatm-ent for pediatric DCM associated with myocarditis, current study shows imp-rovement in patients who received IVIG, this patients may have resent onset dilated cardiomyopathy (which associated with myocarditis). Patients with myocarditis had 3 time's greater rate of recovery and lower rate of death than IDCM (13, 14)

Conclusion: our results revealed that children with DCM of younger age ≤ 12 months, male gender had poor outcome, low SF or EF does not predictor in the outcome. The effectiveness of IVIG therapy in this disorder should be tried in multicenter. In our era DCM needs more study to analyzed the causes, and find ways to early diagnosis and early treatment to avoid heart transplant or early death.

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