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Original Research Article

Clinical course of dilated cardiomyopathy in children

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ABSTRACT

Background: Dilated cardiomyopathy (DCM) in children is a serious disorder. Here authors study the risk factors for predicting prognosis of dilated cardiomyopathy in children.
Methods: An observational case series study done in Institute of child health and research centre, Government Rajaji hospital, Madurai during September 2012 to August 2014. The diagnosis of DCM was based on clinical examination and echocardiographic evidence. Patients were followed up and grouped according to the outcome as improved or cured (group I), no change in clinical status (group II) and worsened or dead (group III).
Results: 31 children less than 12 years of age were diagnosed as DCM. 13 children came under group I with a mean age of 2.15±2.5 and the mean cardiothoracic diameter ratio in chest X-ray was 59.8±3.3. The mean left ventricular ejection fraction at the time of admission was 38.7±3.04 and was 52.1±2.7 on last follow up. 11 children came under Group III with a mean age of 5.35±4.4 and the mean cardiothoracic diameter ratio was 65.3±2.7. The mean left ventricular ejection fraction was 35.8±3.7 at admission and 32.6±2.9 on last follow up. Only two children came under Group II and hence their comparison is negligible.
Conclusions: Children with higher age at the time of diagnosis, higher cardiothoracic diameter ratio in chest X-ray and a low left ventricular ejection fraction on serial echocardiogram were associated with a poor outcome.

Keywords: Cardiomyopathy, Cardiothoracic diameter, Ejection fraction

INTRODUCTION

Cardiomyopathy is the term used for group of diseases of the heart muscle that is not secondary to hypertension, valvular or congenital heart disease or pulmonary vascular disease. Cardiomyopathy is traditionally classified as dilated, hypertrophic or restricted.

Dilated cardiomyopathy refers to a group of condition of diverse etiology in which both ventricles are enlarged with reduced contractility. The known causes of dilated cardiomyopathy include infections, metabolic derangement, hereditary disorders, nutritional deficiencies and exposure to certain drugs and toxins. In spite of the number of etiologies, majority of the patients are classified as idiopathic because the cause is not readily identifiable.

However, there are studies that seem to suggest that viral myocarditis may be the initial insult in many patients.[1] It is difficult to distinguish myocarditis from cardiomyopathy particularly in infants and young children.[2] Very few population based studies have attempted to define the incidence and prevalence of dilated cardiomyopathy in children and risk factors to predict the outcome. A population based study from

Finland by Arola et al estimated the incidence of dilated cardiomyopathy in children to be 0.34/1,00,000/year.[3] At the end of 10 years study period the prevalence was estimated to be 26/1,00,000. Another study from Australia suggested an incidence of 1.09/1,00,000 in children 10 years old or younger for all forms of cardiomyopathies.[4] It has been shown that genetic transmission occurs in 20-30% of patients with dilated cardiomyopathy.[5,6] The actual frequency of dilated cardiomyopathy can be underestimated because, the possibilities of missing affected individuals and when the history of the disease is not evident. Facilities for carnitine assay are limited and the test is very expensive in the developing world. The basis for administration of L-Carnitine to all children with dilated cardiomyopathy irrespective of carnitine levels has been shown to be useful in a multicentric retrospective study involving North American centres and in a small prospective controlled study from India.[7]

The aim is to study the clinical course, analyse the risk factors and predict the prognosis of dilated cardiomyopathy in children admitted in the Institute of child health and research centre, Government Rajaji hospital, Madurai.

METHODS

This was a hospital based study conducted in the Institute of child health and research centre, Government Rajaji hospital, Madurai medical college, Madurai. This observational case series study was conducted during the period of September 2012 to August 2014.

Inclusion criteria

- The diagnosis of dilated cardiomyopathy was based on clinical examination and echocardiographic evidence of systolic ventricular dysfunction. All patients fulfilling the criteria for the diagnosis of dilated cardiomyopathy according to the WHO/ ISFC task force (international society and federation of cardiology) were enrolled in the study.

Exclusion criteria

- Children with Rheumatic heart disease, congenital heart disease, hypertension, Coarctation of aorta, pulmonary vascular disease, Kawasaki's disease, arrhythmia-induced cardiac dysfunction, Takayasu's aortoarteritis, coronary artery abnormalities neuromuscular disease with cardiac involvement and prior exposure to drugs like steroids or anthracyclines were excluded in the study.

The following data were recorded for each patient including age, sex duration of symptoms, preceding history suggestive of viral illness, family history of cardiac illness or sudden death and consanguinity of parents. Detailed clinical examination, chest x-ray, electrocardiograms and serial echo- cardiograms were done for all children. The clinical course treatment and outcomes were noted. Patients were grouped according to the outcome as improved or cured (group I), no change in clinical status (group II) and worsened or dead (group III). Patients who were lost to follow up were not included in these groups.

- Patients with improved clinical status and an increase in the ejection fraction more than 5 % were defined as improved (group I).
- No change or <5% change in the ejection fraction was considered as unchanged (group II).
- Clinical worsening and (or) decline in the ejection fraction >5% were classified as worsened (group III).

Whenever a death was recorded, an attempt was made to ascertain the probable cause of death. Patients who had a follow up for only less than 3 months were classified as lost to follow up.

RESULTS

31 children less than 12 years of age were diagnosed as dilated cardiomyopathy during the period of September 2012 to August 2014 in the Institute of Child Health and Research Centre, Government Rajaji Hospital, Madurai.

Age and sex

In the 31 children diagnosed, their age ranged from 3 month to 12 years. The study group included 9 infants (29%). The mean age of children in present study was 3.5±4.03 years. 24 children in present study group were below 5 years while 7 children were above 5 years. During the study period 5 children were lost to follow up while 10 children died during the study period. In the study group 24 were female children and 7 were male. The age pattern of the cases is noted in Table 1.

Table 1: Age wise pattern of dilated cardiomyopathy in children.

Age in years	No. of cases	Alive	Dead	Lost follow up
0-2 years	19	13	4	2
2-12 years	12	3	6	3
Total	31	16	10	5

Clinical presentation

Congestive heart failure was the initial presentation in nearly 80% of the children. Among them 83% had Grade III or IV dyspnoea (New york Heart association) while the remaining children had Grade II dyspnoea. Two cases were diagnosed as dilated cardiomyopathy while evaluating for the lower respiratory tract infections and one case during evaluation for anaemia These three children had cardiomegaly on radiography, not explainable otherwise.

Family history

No family history suggestive of dilated cardiomyopathy was found in present study group children. One mother of the study children had rheumatic heart disease for which she was taking treatment. But parents of thirteen children (42%) had history of consanguineous marriages. Preceding history of viral fever was present in 9 cases of which six children were below 2 years.

Physical examination

On clinical examination, dysmorphic facial features were not noticed in any of the children. Anaemia was present in eight children and peripheral smear examination revealed dimorphic anaemia in all the cases. Only one child was given packed red blood cell transfusion while other children were supplemented with iron and folic acid and B-Complex vitamins. Hypertension was not seen in any of the children. Grade II systolic murmur was heard in about 11 cases (35%) while the remaining case revealed no murmurs. All these children except the two who were admitted for LRI had gallop rhythm at the time of first presentation.

Chest X- ray

On chest X-ray, cardiomegaly was present in all cases and the mean cardio thoracic ratio was 61.8±3.97.

Electrocardiogram

Electrocardiogram study revealed LVH pattern (Left ventricular hypertrophy) in 59% of children and ST-T changes namely ST elevation and T wave inversion were noticed in 22% children with dilated cardiomyopathy.

Echocardiogram

Echocardiogram study revealed depressed left ventricular systolic function in all the patients. The mean left ventricular ejection fraction was 38±3.8%. An intracavitary thrombus was present in two cases and one child had associated infective endocarditis. Mitral regurgitation was seen in all the children in the first echocardiogram study. Tricuspid regurgitation was seen in nine cases. Serial echocardiograms were taken during follow up of the cases. Improvement in ejection fraction of more than ten percentage was noticed in 13 children while a fall in ejection fraction was noted in three children.

Follow up

Clinical follow up of more than 6 months was available in 19 children (61%). The follow up period ranged from 3 month to 1½ years. All children on follow up received supportive medical treatment with angiotensin converting enzyme inhibitors (Enalapril), digoxin and diuretics. Iron supplementation was given to those children with anaemia. Three children died in present hospital on subsequent admissions after a median follow up of 9-12 months due to uncontrolled congestive cardiac failure. Children were grouped according to the outcome. Those who improved were grouped as Group I, with no change in clinical status as Group II and those who worsened or died as Group III. 13 children came under group I with a mean age of 2.15±2.5. 38% children under Group I had an antecedent history of viral fever. The mean cardiothoracic diameter was 59.8±3.3 in Group I children. The mean left ventricular ejection fraction was 38.7±3.04 at the time of admission while it was 52.1±2.7 on last follow up. The mean age of children in group III was 5.35±4.4 with only 27 % children had an antecedent history of viral fever. The mean cardiothoracic radio was 65.3±2.7 in Group III children. The mean left ventricular ejection fraction was 35.8±3.7 at the time of admission and 32.6±2.9 on last follow up. Group II children were only two in number and hence their comparison is negligible. Both were females and aged 1-2 years. The clinical and laboratory variables of the groups are given in Table 2.

Table 2: Clinical and laboratory variables in different subgroup of children.

Variables	Improved Group I	No change Group II	Worsened/ died Group III	P value
No. of children	13	2	11	NA
Age	2.15±2.5	1.12±0.12	5.35±4.4	<0.001
Male/Female	3:10	0:2	3:8	NA
Antecedent viral fever	38%	-	27%	NA
Cardiothroracic ratio	59.8±3.3	61±1.1	65.3±2.7	<0.001
Left ventricular hypertrophy	31%	50%	81%	NA
ST-T changes in ECG	21%	50%	36%	NA
LVEF* at admission	38.7±3.04	38±0.5	35.8±3.7	<0.05
LVEF* at last follow up	52.1±2.7	35±0.5	32.6±2.9	<0.001

*Left ventricular ejection fraction

DISCUSSION

In present study the number of females affected were more than males in all age groups. In all other studies this sex prevalence is not evident. This differential incidence in sex could not be explained and this requires further study to confirm that if there is an actual increase in female incidence.

On univariate analysis gender, symptom duration, New York Heart Association class dyspnoea at presentation, history of antecedent viral infection, left ventricular hypertrophy or ST-T changes on electro cardiogram did not predict the outcome. Only higher the age at diagnosis, higher cardiothoracic ratio and a low left ventricular ejection fraction on serial echocardiogram were associated with a poor outcome.

The mean age of children who improved was 2.15±2.5, as against the mean age of children who worsened or died was 5.35±4.4. The mean cardiothoracic ratio was 59.8±3.3 in those who improved while it was 65.3±2.7 in those who worsened or died during the follow up. There was an increase in left ventricular ejection fraction (14.6±2.9) in the improved group of children on serial follow up echocardiograms. A fall in the left ventricular ejection fraction (3.2±2.45) was noted in the Group III children. Due to the small absolute number of events and patients, a multivariate analysis of predictive variables was not done.

Several studies in the past have addressed the issue of the natural history of dilated cardiomyopathy in children and the predictive variables influencing the clinical course of the disease. In support of present study, better prognosis in infants compared to children beyond infancy was reported in few studies.[8-11] Infants are expected to have higher regeneration capabilities then older children.[12] Higher incidence of dilated Cardiomyopathy is noted in infants in present study and also in AIIMS, Australian, and American studies and better prognosis in infants was observed in American study.[13-15] Other studies also have reported poor prognosis in older children. But AIIMS study has reported poor prognosis in infants.

Familial dilated cardiomyopathy has been reported in nearly 20% of patients reported from western countries. But no single case was found in present study. Sudden death is reportedly rare in children with dilated cardiomyopathy compared to adults but has been documented in 3.6% children in one Australian study.[16] Present study did not note any sudden death.

CONCLUSION

From the multitude of investigations and treatment strategies that are recommended for dilated cardiomyopathy, it is clear that the etiopathogenesis of paediatric dilated cardiomyopathy is poorly understood. Higher age at diagnosis was associated with poor prognosis in present study. Higher cardiothoracic ratio in X-ray chest and persistent low left ventricular ejection fraction values at serial echocardiogram were associated with poor prognosis.Treatment of Paediatric dilated cardiomyopathy continues to be largely supportive in present country and prognosis may not improve without further understanding of its etiologies and development of etiology-specific therapies.

Recommendations

The clinical features of dilated cardiomyopathy are mainly nonspecific and high index of suspicion is needed to identify the cause. Long term follow up of children with dilated cardiomyopathy is necessary to arrive clearly at the predictors of prognosis. Sophisticated laboratory work up is needed to arrive at etiological diagnosis.

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