

FREQUENCY & ECHOCARDIOGRAPHIC STUDY OF DILATED CARDIOMYOPATHY IN CHILDREN PRESENTING WITH CARDIAC FAILURE

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ABSTRACT

Objective: To evaluate the role of echocardiography in diagnosis of dilated cardiomyopathy as a cause of cardiac failure in children.

Design: This was descriptive study. Children presenting with cardiac failure from indoor patients were selected and echocardiography along with chest X- ray, ECG, cardiac enzymes & ASO titre was performed in all patients.

Setting: Hospitalized patients included in the study were selected from the Department of Child Health, Khyber Teaching Hospital.

Subject: Fifty hospitalized patients with congestive heart failure were selected consecutively from hospitalized patients.

Main Outcome: Role of echocardiography in the diagnosis of dilated cardiomyopathy in children presenting with cardiac failure.

Results: Out of fifty patients admitted with cardiac failure 27 (54%) cases were found to be dilated cardiomyopathy while congenital heart disease, myocarditis & rheumatic heart disease were found in 12 (24%), 8 (16%) & 3 (6%) cases respectively.

Conclusion: Dilated cardiomyopathy is an important cause of cardiac failure in children & echocardiography is an important tool to diagnose and differentiate dilated cardiomyopathy from other causes of cardiac failure.

KEY WORDS: Cardiac failure, dilated cardiomyopathy, echocardiography

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INTRODUCTION

The cardiomyopathies are defined as diseases of the myocardium associated with cardiac dysfunction.¹ The term "cardiomyopathy" is used to describe a clinical syndrome in which abnormality seems to reside in the myocardium itself.²

Cardiomyopathy is regarded as primary when the heart is considered to be the only organ involved. In secondary cardiomyopathy heart lesion is part of a systemic disease or has a known cause.²

Cardiomyopathies have recently been classified into following groups:¹

1. Dilated cardiomyopathy
2. Hypertrophic cardiomyopathy

3. Restrictive (Infiltrative) cardiomyopathy
4. Arrhythmogenic right ventricular Dysplasia / cardiomyopathy.
5. Specific cardiomyopathies

DILATED CARDIOMYOPATHY

Dilated cardiomyopathy (DCM) is a syndrome characterized by cardiac enlargement and impaired systolic function of one or both ventricles.³ Mostly left ventricular and also right ventricular contractility is diminished, leading to decrease in cardiac output and increased end systolic volumes and pressures.⁴ While formerly it was called congestive cardiomyopathy, the term dilated cardiomyopathy is now preferred, since the earliest abnormality usually is ventricular enlargement and systolic contractile dysfunction, with congestive heart failure often (but not invariably) developing later. Idiopathic dilated cardiomyopathy leads to symptomatic congestive heart failure (CHF) in majority of patients and carries a high morbidity and mortality.⁵

The reported annual incidence of the idiopathic dilated cardiomyopathy in the United States is between 5-8 cases per 100,000 persons^{4,6} and dilated cardiomyopathy is the cause of congestive heart failure (CHF) in approximately 25% of all cases of CHF.⁷ The disease follows a seasonal pattern, that is a much larger number of acute cases are seen in months of early Spring (February, March) and Winter (October, November).⁸

The natural history of idiopathic DCM varies widely. Although case fatality rates approach 30% at 3 years and 60% at 5 years, complete recovery of left ventricular (LV) function may also occur.⁹

Dilated cardiomyopathy is a leading cause of heart failure, with a prevalence of 36.5 per 100,000 individuals. Several studies indicate that 25% to 30% of DCM is familial.¹⁰

PATIENTS AND METHODS

This study was conducted in the Paediatric Department of Khyber Teaching Hospital,

Peshawar. The descriptive study included 50 patients who were hospitalized with signs and symptoms of congestive heart failure, during 8 months period from October 2001 to May 2002. Inclusion criteria was age ≤ 15 years with signs and symptoms of congestive heart failure. All patients above 15 years or having cause other than heart pathology were excluded from the study.

A detailed medical history was taken (including age, gender, presenting complaints, consanguinity, family history, past history) from all the patients fulfilling the inclusion criteria and complete clinical examination (including general physical examination and systemic examination especially the cardiovascular system) was carried out.

The clinical features of congestive cardiac failure were oedema (periorbital, pedal, sacral), tachycardia, tachypnea, gallop rhythm, cardiomegaly, hepatomegaly and basal crepitations.

The diagnosis of dilated cardiomyopathy was confirmed and differentiated from other causes of CHF on the basis of history, physical examination, roentgenogram of the chest, ECG and echocardiography. SPSS was used for data analysis. Mean, percentage and standard deviation were used for analyzing the whole data and documenting the final result.

RESULTS

Fifty cases of congestive heart failure were studied from October 2001 to May 2002, admitted to Paediatric Department, Khyber Teaching Hospital. The study showed dilated cardiomyopathy as the most common cause of congestive heart failure in paediatric age group constituting about 54% of the total cases, followed by congenital heart disease, myocarditis and rheumatic heart disease constituting 24%, 16% and 6% respectively.

Among 27 cases of dilated cardiomyopathy 18 (66.7%) were males and 9 (33.3%) were females. The mean age was 5.07 ± 3.46 years in male patients and mean age in case of female patients was 5.11 ± 3.54 years.

TABLE-I
Frequency of DCM in different
paediatric ages

Age (years)	No of patients	Percentage
< 1 Year	2	7.4%
2—5	17	63.0%
6—10	6	22.2%
11—15	2	7.4%

Congenital heart disease was common in infantile age, rheumatic heart disease was common above 8 years while myocarditis was common in almost the same age group as it was in the case of dilated cardiomyopathy i.e all age groups but most common in age group two to three years. (Table-I)

The most common presenting complaints were dyspnoea, oedema, tachycardia, tachypnoea present in almost all patients while pallor, fatigue and irritability found in about 70%. (Table-II)

Chest radiograph showed cardiomegaly in all cases (100%) with marked cardiomegaly in 70% of cases. Electrocardiography revealed tachycardia in all cases (100%).

The common features of dilated cardiomyopathy on echocardiography included left ventricular global hypokinesia, systolic dysfunction, markedly reduced fractional shortening, pulmonary hypertension, regurgitant valvular lesion (MR, TR) and rarely pericardial effusion. The mean left ventricular end-diastolic diameter was 5.06 ± 1.01 cm, left ventricular end-systolic diameter was 4.23 ± 0.91 cm, ejection fraction $32.63 \pm 9.8\%$ and fractional shortening $16.3 \pm 5.02\%$ as given in table-III.

In this study six patients had positive family history constituting 22.22% of all cases.

DISCUSSION

This study showed dilated cardiomyopathy as the commonest cause of heart failure in children (≤ 15 years). Other causes of heart failure were congenital heart disease, myocarditis and rheumatic heart disease respectively.

TABLE-II
Clinical features of patients
with DCM

Age (In years)	No. of Patients	Dys pnoea	Oede ma	Cough	pallor	Hepato megaly	Tachy cardia
<1 Year	2	+	+	+	+	+	+
2-5	17	+	+	+	+	+	+
6-10	6	+	+	+	--	+	+
11-15	2	+	+	+	--	+	+
Total	27	100	100	100	70.4	100	100

TABLE -III
Echocardiographic findings in patient
with DCM

Cardiac Parameter	Mean	Standard Deviation
LVEDD	5.05	± 1.01
LVESD	4.23	± 0.91
FS (%)	16.3	± 5.02
EF (%)	32.63	± 9.8

LVEDD : Left ventricular end diastolic diameter
LVESD : Left ventricular end systolic diameter
FS : Fractional Shortening
EF : Ejection Fraction
DCM : Dilated cardiomyopathy

The frequency of dilated cardiomyopathy is comparable with national reported figures.¹¹

In this study congenital heart disease was common below 2 years. Rheumatic heart disease was found from 6 to 15 years and dilated cardiomyopathy and myocarditis presented in all paediatric ages (upto 15 years) but commonly between 2 to 5 years. The incidence of dilated cardiomyopathy was least common below the age of one year and above 10 years of age i.e 7.4%. A hospital based study from Karachi showed that acquired heart diseases are most common above six years and least common in first year of life.¹² The number of cases were more during Winter and early spring. This is consistent with a study carried out at Lahore by Sadiq M et al according to which the disease follows a seasonal pattern. A much larger number of acute cases are seen in months of early Spring (February, March)

and Winter (October, November).⁸

The high incidence of dilated cardiomyopathy is because of unawareness of this condition in the medical community. The patients are not brought to hospital in time because they mostly present with shortness of breath and pallor, unfortunately treated in periphery as cases of respiratory tract infection or anaemia.

The Chest roentgenogram revealed marked cardiomegaly in about 70% while mild to moderate cardiomegaly in rest of the cases. Pleonemic lung fields were common while pulmonary oedema was found rarely.

Electrocardiography showed tachycardia in all cases (100%), low voltage waves and complexes and ST segment and T-wave changes in 10 cases (37%).

The Echocardiographic study revealed marked left ventricular dilatation and generalized left ventricular hypokinesia in almost all patients with dilated cardiomyopathy. The same was true for systolic dysfunction. Other important echocardiographic features were mitral valve regurgitation, tricuspid valve regurgitation and pulmonary hypertension in about 22.22% of patients. The mean fractional shortening was $16.3 \pm 5.02\%$ and ejection fraction was $32.63 \pm 9.8\%$. Left ventricular end-diastolic and end-systolic diameters were 5.05 ± 1.01 cm & 4.23 ± 0.91 cm respectively. These results are closely consistent with the results documented in a study by Rehan Ahmad and Zahid Awan.¹¹

The familial incidence of dilated cardiomyopathy was 22.22% which is closely consistent with the international figures(Tired L et al and Grunig E et al).^{13,14}

Common complications associated with dilated cardiomyopathy were arrhythmias, ventricular clots, bacterial endocarditis and stroke (secondary to thromboembolism).

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