

CARDIOMYOPATHIES: PRESENTATION AND COMPLICATIONS IN CHILDREN AT DERA ISMAIL KHAN

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ABSTRACT

Background: Cardiomyopathies are a group of non-inflammatory conditions of myocardium leading to cardiac dysfunction. This study was conducted to see the presentation and complications of cardiomyopathies in children at Dera Ismail Khan.

Material & Methods: This descriptive case series was carried out at Department of Paediatrics, DHQ Teaching Hospital, Dera Ismail Khan, from January 2007 to December 2008. All cases admitted with congestive heart failure were investigated. After history and examination, investigations including CXR, ECG and Echocardiography were performed. Other relevant investigations were also performed as and when indicated. Both already and newly diagnosed cases were included in the study. Exclusion criteria were cause of CCF other than cardiomyopathy and/or a normal echocardiography. Data was analyzed for age, sex, presentation, complications, associated conditions, and mortality.

Results: Sixty patients were admitted with cardiomyopathy during the study period. All these patients initially presented with congestive cardiac failure. Sixteen (27.6%) of these were being treated as wheezy bronchitis/asthma by local doctors. Commonest type was dilated CMP 54(90%). Other less common types were hypertrophic CMP 4(6.6%) and restrictive CMP 2(3.3%). Common complications included failure to thrive 35(58.3%), mitral regurgitation 8(13.3%), pericardial effusion 7(11.7%), hemiplegia 5(8.4%), pulmonary hypertension 4(6.7%), arrhythmias 2(3.4%), pleural effusion 2(3.4%), infective endocarditis 1(1.1%), shock 1(1.1%) and death 1(1.1%).

Conclusion: Commonest presentation of cardiomyopathy is congestive cardiac failure, but repeated wheezy attacks are also common during early stages of the disease. Commonest complications are mitral regurgitation, pericardial effusion, stroke and failure to thrive.

Key words: Cardiomyopathy, Congestive cardiac failure, Wheezy bronchitis.

INTRODUCTION

Cardiomyopathies (CMP) are a group of non-inflammatory conditions of myocardium leading to cardiac dysfunction.¹⁻³ These may be primary or idiopathic and secondary due to endocrine disorders, metabolic diseases, nutritional derangements, neuromuscular disorders, blood diseases, genetic disorders, tumors, mitochondrial disorders and arrhythmias.^{1,4,5} Based on predominant structural and functional abnormalities, recent classification is as under:

1. Dilated or Congestive Cardiomyopathy
2. Hypertrophic Cardiomyopathy (HCM)
3. Restrictive Cardiomyopathy
4. Arrhythmogenic right ventricular dysplasia
5. CMP secondary to systemic diseases^{1,2,6-8}

Prevalence of CMP in children is 36/100,000 for dilated Cardiomyopathy (DCM) and 2/100,000

for hypertrophic and restrictive CMP.¹ Usual presentation is with heart failure and cardiomegaly on chest X-ray.⁹ Other features include failure to thrive, recurrent chest infections and easy fatigueability. In younger children, abdominal symptoms may be prominent.¹ Treatment is type specific and includes supportive measures (dietary adjustments, anti-failure measures), drugs (ACE inhibitors, β -blockers, anti-arrhythmic drugs, anticoagulation), implantable cardiac defibrillator, immuno-suppression and surgery (heart transplantation).^{1,10}

Present study was conducted to see the types, presentation and complications of cardiomyopathies in children.

MATERIAL AND METHODS

This descriptive case series was conducted in the Department of Paediatrics, DHQ Teaching Hospital attached to Gomal Medical College, Dera

Ismail Khan. Study period was 2 years; January 2007 to December 2008.

All those patients who were admitted with symptoms and signs of congestive heart failure were included in the study. A detailed medical history was taken including presenting complaints, past history and family history. A complete clinical examination was performed particularly for heart disease and its complications. Clinical features of congestive heart failure considered were tachycardia, tachypnea, orthopnea, edema, gallop rhythm, hepatomegaly, cardiomegaly, and basal crepitations in various combinations. Any other features including complications were also noted.

Patients were investigated with x-ray chest, ECG and echocardiography. Other relevant investigations (FBC, MP, ALT, urinalysis, BUN, creatinine, CPK, cultures, ultrasonography of abdomen/chest) were also performed when indicated. Patients were also investigated for complications of CMP (CT scan, MRI, USG abdomen/chest).

The diagnosis of dilated CMP was based on cardiomegaly on x-ray chest and decreased left ventricular systolic function on echocardiography; of HCM on disproportionate hypertrophy of left ventricle (especially septum) on echocardiography; and of restrictive CMP on decreased diastolic dilatation of left ventricle on echo-cardiography.

Both already diagnosed and newly diagnosed cases of CMP were included in the study. Exclusion criteria were causes of CCF other than CMP and/or normal echocardiography.

Data was analyzed for age, sex, presentation, complications, associated conditions and mortality.

RESULTS

Sixty patients were diagnosed as cases of cardiomyopathy during the study period. All these patients belonged to D.I.Khan District. Median age at the time of presentation was 3.9 years with youngest patient being 3 months old and eldest 12 years. Males were 33 (55%) and females 27 (45%). (Table-1)

Table 1: Age and sex distribution of patients.

Age Sex	<1 year	1-5 years	5-12 years	Total
Male	6	16	11	33
Female	5	18	4	27
Total	11	34	15	60

All the 60 patients initially presented with congestive cardiac failure but 16 (27.6%) of them were being treated as wheezy bronchitis /asthma by local doctors without having an x-ray chest.

Various types of CMP seen were dilated CMP 54 (90%), HCM 4 (6.6%), and restrictive CMP 2 (3.4%).

Complications included poor growth in 35 (58.3%) children, mitral regurgitation in 8 (13.3%), pericardial effusion in 7 (11.7%), hemiplegia in 5 (8.4%), pulmonary hypertension in 4 (6.7%), arrhythmias (atrial fibrillation) in 2 (3.4%), pleural effusion 2 (3.4%), infective endocarditis 1 (1.7%), shock 1 (1.7%), and 1 patient (1.7%) died indoor during the study period. (Table-2) Three patients were observed to be cured completely after 6-12 months of follow-up.

Table 2: Complications in patients with cardiomyopathy.

S. No.	Complications	Number (%)
1.	Failure to thrive	35 (58.3%)
2.	Mitral regurgitation	8 (13.3%)
3.	Pericardial effusion	7 (11.7%)
4.	Hemiplegia/stroke	5 (8.4%)
5.	Pulmonary hypertension	4 (6.7%)
6.	Arrhythmias (atrial fibrillation)	2 (3.4%)
7.	Pleural effusion	2 (3.4%)
8.	Infective endocarditis	1 (1.7%)
9.	Shock	1 (1.7%)
10.	Death (due to AF in HCM)	1 (1.7%)

Etiology could not be confirmed in majority of cases but in a few, there was some clue to the cause. These included post-viral myocarditis, diphtheria and Gorgoylism (Hurler syndrome). (Table-3)

Table 3: Etiology /Associated conditions.

S. No.	Cause /Associated condition	Number (%)
1.	Post-viral myocarditis	3 (5.0%)
2.	Diphtheria	1 (1.7%)
3.	Gorgoylism (Hurler syndrome)	1 (1.7%)
4.	Acute viral hepatitis	1 (1.7%)
5.	Chronic renal failure	1 (1.7%)
6.	Ricketts	1 (1.7%)

DISCUSSION

Cardiomyopathies are a heterogenous group of relatively uncommon chronic conditions in pediatric practice, but have high morbidity and mortality. This increases the importance of these conditions several folds.^{1,11} These patients need repeated hospitalizations for recurrent chest infections and attacks of congestive heart failure; thus the burden on the hospitals is more than the actual number of patients.

No age is immune but in our study median age (3.9 years) at diagnosis was somewhat higher than some studies^{9,12} but comparable to others.² This may be a geographical variation.

All the patients in our series presented with congestive cardiac failure but a significant number of patients (27.6%) were being treated as wheezy bronchitis and asthma by local doctors without having an x-ray chest. The diagnosis was only suspected and later confirmed after having and x-ray chest. This misconception is not uncommon and has been reported by other workers as well.² It is therefore advisable to have an x-ray chest for recurrent wheezy attacks before labeling a patient as asthmatic.

In our study, 90% of the patients were suffering from dilated CMP. Same figures are reported by other workers as well,^{2,13} but lower figure of 10% is reported from Karachi.¹⁴ This difference may be due to geographical and seasonal variation or a bias of a retrospective study.

The commonest complication in our series was mitral regurgitation (13.3%). All the patients were suffering from DCM. It is well known complication and occurs due to dilatation of valvular ring. It is a contributory factor towards the development of pulmonary hypertension.² Next common complication was pericardial effusion (11.7%). This is probably common in post viral myocarditis and has been reported by other workers also.⁹ A significant complication in our study was hemiplegia (8.4%) due to thrombo-embolic phenomenon. Its frequency in our study supports the regular long term use of anti-coagulation.

Most of the complications in our patients were within the heart and chest but 6 patients had extra-thoracic complications including stroke and shock. One patient died during hospitalization due to atrial fibrillation. This patient was suffering from HCM, which is a well known cause of arrhythmias and sudden death.^{15,16}

Although CMP is a well known complication of Duchenne and other muscular dystrophies,^{6,8} none of our patients was suffering from muscular dystrophy. The reason is not clear, but it may be a

chance or the rarity of muscular dystrophies in this area.

As expected, etiology could not be established in the majority of cases, but in 3 cases echocardiographic diagnosis was post-viral myocarditis, one case was post-diphtheria and one case was due to gorgoylism (Hurler syndrome). All of these are well known causes of CMP.¹ Other associated conditions found during the study included acute viral hepatitis, chronic renal failure and rickets in one case each. The role of these conditions in causation and pathogenesis of CMP, if any, is not known. These conditions might have been coincident findings.

CONCLUSIONS

Commonest presentation of cardiomyopathy is congestive cardiac failure, but repeated wheezy attacks are also common during early stages of the disease. Commonest complications are mitral regurgitation, pericardial effusion, stroke and failure to thrive.

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