

## Echomorphology of cardiomyopathy: review of 217 cases from 1999 to 2010

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### Abstract

**Objective:** To study echocardiogram features of different types of cardiomyopathy presenting over a 12 year period at a single centre in Peshawar.

**Methods:** The series comprised a retrospective review of 13,788 consecutive echocardiograms carried out at the Muhammadi Hospital International Medical Research Centre, Hayatabad, Peshawar, from January 1999 to December 2010. Patients were split into two: Group I with paediatric and adolescent cases (0-18 years) and Group II with adults (>18 years). In the adult group, women with peripartum cardiomyopathy were subdivided into two groups of 18-30 years and 30 to 44 years. Standard Echo B and M modes and Doppler parameters were recorded to ascertain the diagnoses of common primary and secondary cardiomyopathies. Patients with myocarditis with chambers' dilatation and global dysfunction, and cardiopathy associated with major cardiovascular diseases were excluded. SPSS 14 was used for statistical analysis.

**Results:** Cardiomyopathy was diagnosed in 217 (1.57%) cases. There were 144 (66%) cases of dilated cardiomyopathy with a mean age of 13±14.8 years; 17 (8%) cases of hypertrophic cardiomyopathy with a mean age of 12±11.5 years; and 7 (3%) cases of restrictive cardiomyopathy with a mean age of 31±7.8 years. Primary cardiac amyloidosis was confirmed in 9 (4%) cases, and peripartum cardiomyopathy in 25 (11%) females. Rare subtypes were found in 15 (7%) cases.

**Conclusion:** DCM was the most frequently diagnosed subtype of cardiomyopathy followed by HCM in both the adult and paediatric age groups.

**Keywords:** Congestive cardiac failure, Myocarditis, Cardiopathy, Myocardiopathy. (JPMA 63: 454; 2013)

### Introduction

The incidence of cardiomyopathy, a heterogeneous group of myocardial disorder, is rising globally, partly due to increased awareness and improved diagnostic techniques. Conflicting definitions and classifications, over the past half-a-century have continued to deprive physicians of the confidence to diagnose. Thus, its clinical impact has remained shrouded with ambiguity. Cardiomyopathies are currently classified as primary with predominant involvement of the heart, and secondary with myocardial involvement due to multi-organ disease.<sup>1</sup> Subtypes in the primary group include dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), restrictive cardiomyopathy (RCM) and peripartum cardiomyopathy (PCM). Common secondary cardiomyopathies include endomyocardial fibroelastosis (EFE), hypereosinophilic syndrome or Loffler's endocarditis, cardiac amyloidosis, systemic lupus

erythematosus (SLE), scleroderma, neuromuscular dystrophy, doxorubicin toxicity and radiation. In contrast, myocardial structural and functional disorders in congenital, rheumatic, ischaemic or hypertensive heart disease, causing congestive cardiac failure,<sup>2</sup> may be aptly called 'cardiopathy,' such as ischaemic cardiopathy.

Myocarditis is the commonest, and cardiomyopathy is the second commonest cause of cardiac failure in children in our country.<sup>3</sup> DCM has been reported to be the commonest type of cardiomyopathy in Pakistan,<sup>4-6</sup> and HCM has emerged as a formidable challenge.<sup>7-9</sup> PCM, a relatively newer entity, had also been implicitly reported as a force to reckon with in our country.<sup>10-12</sup>

The current study was planned to evaluate echomorphology of the prototypes of cardiomyopathy in our community, to highlight their relative presence, which hitherto have not been liberally labelled as such, partly due to hesitation on the part of physicians owing to criterion dilemma. The global diagnostic criteria for cardiomyopathy were used for gross echo-morphological and dynamic features of the various sub-types. This collaborative effort also aimed at increasing the index of suspicion by the medical profession, and to promote awareness in the community about this lingering ailment which requires long-term management.

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## Patients and Methods

The cross-sectional study was done at the Muhammadi Hospital International Medical Research Centre (MH-IMRC) in Hayatabad, Peshawar. A series of 13,788 consecutive echocardiograms recorded between January 1999 and December 2010 were reviewed. Permission from the institution's ethics committee was obtained for review. The sample size was calculated with the World Health Organisation (WHO) sample size calculator formula  $n = Z^2 * P (1-P) / \Delta^2$ .

Keeping 23.9% prevalence at 95% confidence interval and a precision of 5%, the calculated sample size is 217. Non-probability consecutive sampling was employed. To control the bias, each finding was verified by two doctors. Inclusion criteria was children and adults with heart-related complaints and cardiovascular disease. Referrals came from the Children Unit-A, Khyber Teaching Hospital, Children Unit, Naseer Teaching Hospital, the Afghan Polyclinic Peshawar, and the Paediatric Clinic, Kohat. Several cases were followed up more than once, and siblings in a few families were also screened for cardiomyopathy.

Exclusion criteria was based on cases of myocarditis with chambers' dilatation and global dysfunction, and cardiopathy associated with major cardiovascular diseases.

Patients were split into two categories: Group I with paediatric and adolescent cases (0-18 years) and Group II with adults (>18 years). In the adult group, women with peripartum cardiomyopathy were subdivided into two groups of 18-30 years and 30 to 44 years.

Chest x-rays and electrocardiograms were recorded in all these cases. Echocardiograms were obtained using a Siemens Sonoline-SI 450 echocardiograph, and all atypical cases were videographed by a Panasonic SVHS recorder, and diagnostic video prints were obtained by using a Mitsubishi VCP. Two dimensional (B-mode), M-mode and pwD and cwD data, including chamber geometry and myocardial thickness, ejection fraction (EF), fractional shortening (FS) and early/arterial (E/A) ventricular filling velocity ratio were recorded. The diagnosis of cardiomyopathy was made according to the definitions and classification of cardiomyopathies.<sup>1</sup>

DCM was diagnosed by two dimensional B-mode, showing left ventricular spherical dilatation, and lateral and apical displacement of the papillary muscles, causing mitral regurgitation. Four chambers' enlargement with global hypokinesia and myocardial thinning were the diagnostic hallmarks. HCM demonstrated left ventricular hypertrophy, with more marked asymmetrical inter-ventricular septal (proximal) hypertrophy. M-mode showed dynamic left ventricular outflow tract (LVOT) obstruction during systole,

with increased late-systolic gradient after exercise on Doppler study. RCM was diagnosed with biatrial dilatation, decreased left ventricular compliance (delayed relaxation) with diastolic dysfunction. Left ventricular size remained normal, but at the end-stage hypertrophy and systolic dysfunction developed.

PCM generally showed features akin to DCM, including left ventricular dilatation with global hypokinesia. PCA on the B-mode showed left ventricular hypertrophy with valvular involvement, and increased ventricular speckled texture. In EFE, endomyocardial thickening resulted in apical obliteration and restrictive left ventricular compliance with diastolic dysfunction. In arrhythmogenic right ventricular cardiomyopathy (ARVC), the dysplastic right ventricle had segmental dilatation, with regional or global systolic dysfunction, and ventricular arrhythmias. Left ventricular noncompaction (LVNC) showed spongy echo appearance of the left ventricular cavity, particularly in the apical area. It is considered to be due to deficient trabecular formation from incomplete morphogenesis.

The data obtained was analysed on SPSS 14. Descriptive statistics were calculated for each group. Mean  $\pm$  standard deviation (SD) and ranges were calculated for age in years. Percentages were calculated for the types of groups. The male-to-female ratio was also recorded.

## Results

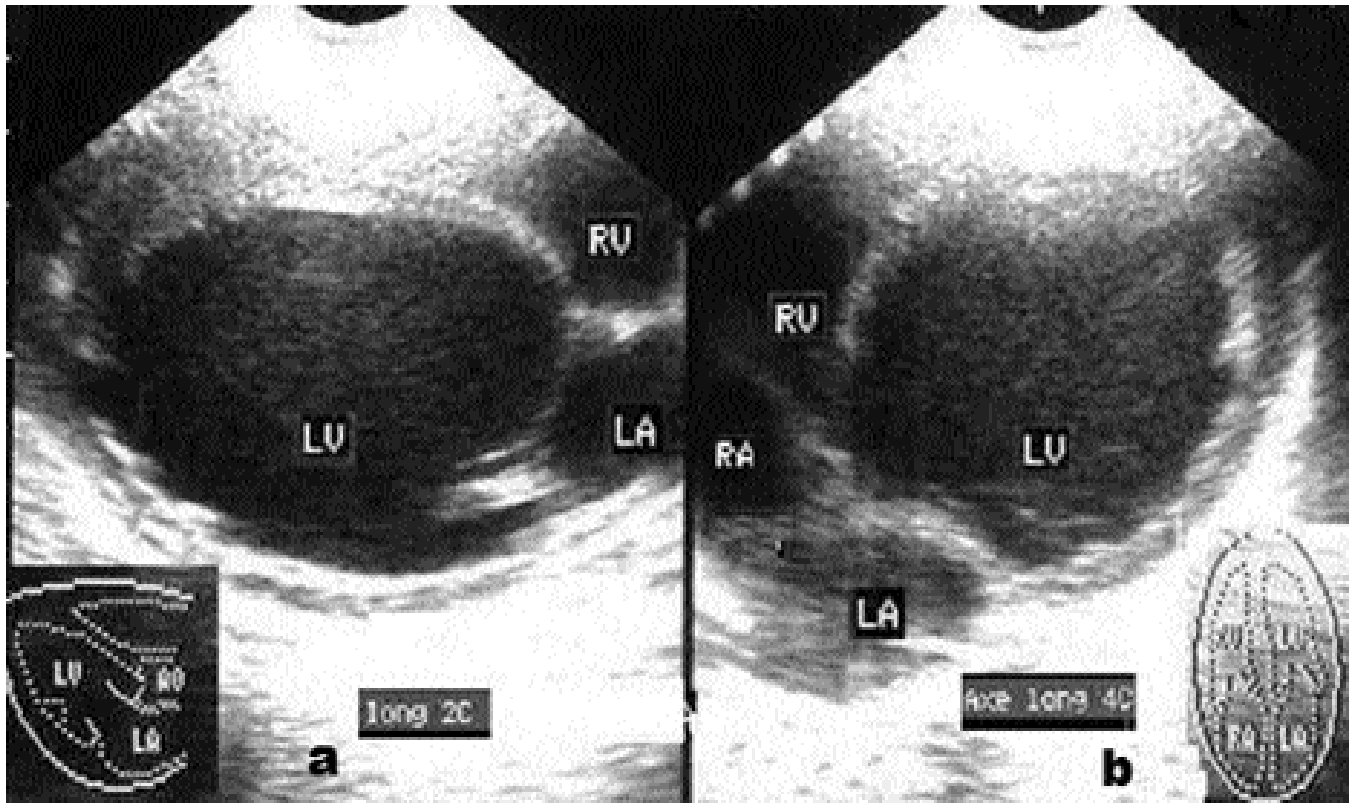
From a series of 13,778 consecutive echocardiograms, 217 cases of cardiomyopathy were located to meet sample size requirements. DCM was present in 144 (66%) cases whose mean age was  $13 \pm 14.8$  years. In Group I, there were 105 cases of DCM [69 (65.7%) boys and 36 (34.3%) girls] with mean age of  $4 \pm 3.9$  years. In Group II, DCM was diagnosed in 39 cases [24 (61.5%) males and 15 (38.5%) females] with mean age of mean  $33 \pm 17$  years (Figure-1).

HCM was present in 17 (8%) cases with a mean age of  $12 \pm 11.5$  years. In Group I, there were 13 (76%) cases of HCM with mean age  $5 \pm 5.9$  years. They included 9 (69.2%) boys and 4 (30.8%) girls. In Group II, there were 4 (24%) HCM cases with mean age  $26 \pm 6.1$  years. They included 3 (75%) males and 1 (25%) female.

RCM was recorded in Group II only: there were 7 (3%) cases with the mean age of  $31 \pm 7.8$  years. There were 3 (42.85%) males and 4 (57.14%) females.

PCA was confirmed in 9 (4%) cases: five males in Group I (mean age  $4 \pm 4.4$  years); and four cases in Group II.

PCM was diagnosed in 25 (11%) females 20-45 years of age (mean:  $35 \pm 8.6$ ). Cases of rare subtypes ( $n=15$ ; 7%), included EFE 3 (20%), LVNC 3 (20%), AVRC 2 (13.33%), scleroderma 3 (20%), SLE 1 (6.66%), and muscular-

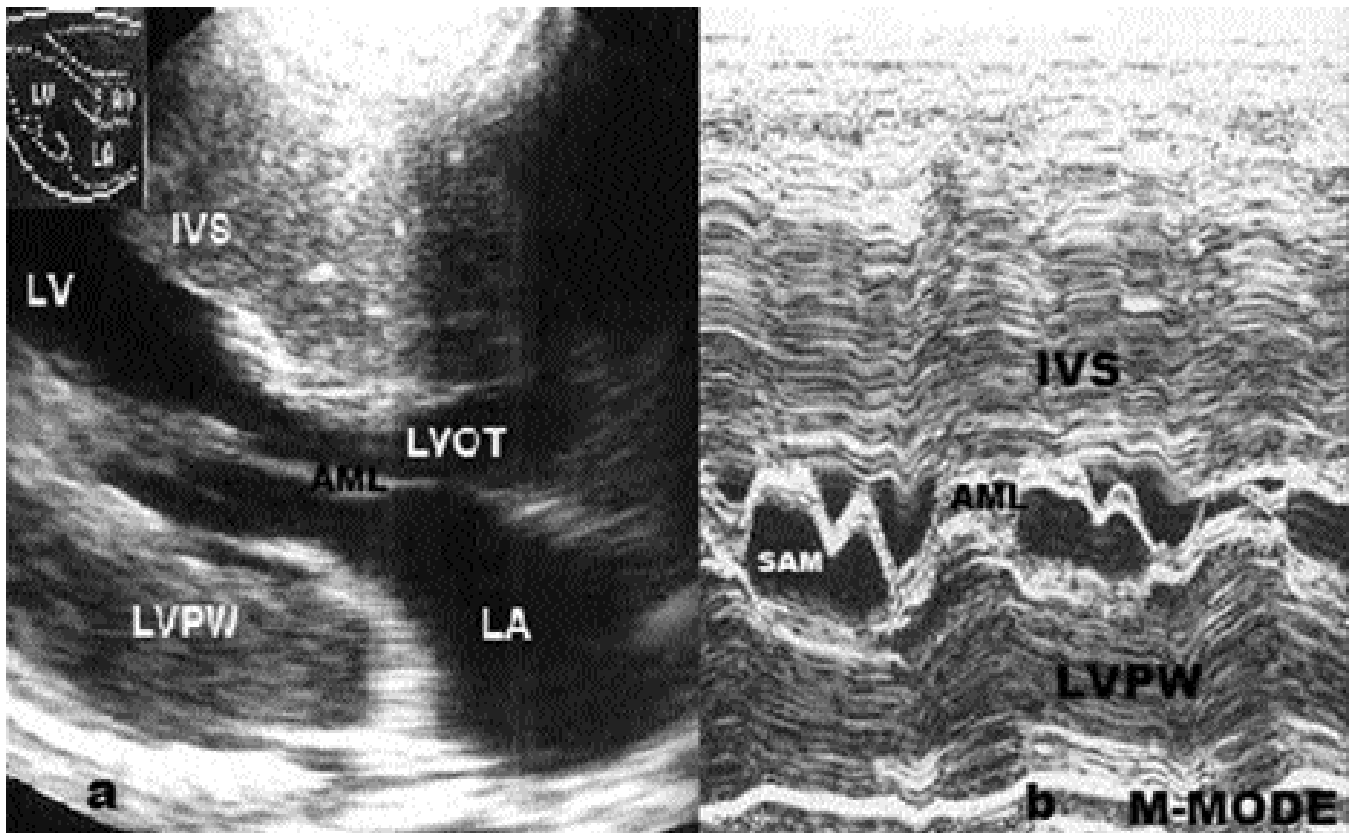


**Figure-1:** I.B: Four-year-old girl; spherically dilated left ventricle in dilated cardiomyopathy (DCM), in two dimensional (B-mode): a. Two chambers long axis view (2CLX), b. Four chambers long axis view (4CLX).

Table: Echo-based diagnostic profile of cardiomyopathy in 217 cases.

	<b>Cardiomyopathies</b>	<b>Total cases</b>	<b>Age range (years)</b>	<b>Mean age (years)</b>	<b>Male:Female Ratio</b>	
1.	Dilated cardiomyopathy (DCM)	144	1-50	13±14.8	2:1	
		105	Group I (1-18)	4±3.9	3:1	
		39	Group II (>18)	33±17	2:1	
2.	Hypertrophic (HCM)	17	1-33	12±11.5	3:1	
		13	Group I (1-18)	5±5.9	2:1	
		4	Group II (18-33)	26±6.1	3:1	
3.	Restrictive (RCM)	7	13-43	31±7.8	3:4	
		2	13-15	14±8.1	1:1	
		5	25-33	28±3.5	2:3	
4.	Primary cardiac amyloidosis (PCA)	9	1-47	15±13.8	7:2	
		5	1-9	4±4.4	4:1	
		4	18-36	29±7.8	3:1	
5.	Peripartum (PCM)	25	20-43	35±8.6	—	
		11	20-30	27±3.8	—	
		14	31-44	38±3.7	—	
6.	Rare (Subtypes)	15	5-28	17±12	—	
		EFE†	3	3-14	9±6	—
		LVNC*	3	23-28	25±3	—
		ARVC#	2	2-3	2.5±1	—
		Others**	7	2-28	14±12	—

†EFE: Endomyocardial fibroelastosis, \*LVNC: Left ventricular noncompaction, #ARVC: Arrhythmogenic right ventricular cardiomyopathy, \*\*Others: muscular- dystrophic cardiomyopathy (3), scleroderma (3), and Systemic lupus erythematosus (SLE) (1).



**Figure-2:** Thirteen-year-old boy, hypertrophic cardiomyopathy (HCM): a. Marked intraventricular septal (IVS) and left ventricular posterior wall (LVPW) hypertrophy in long axis two chambers view (2CLX), with critical left ventricular outflow tract obstruction (LVOT) during systole, b. Septal (IVS) and left ventricular posterior wall (LVPW) hypertrophy, with systolic anterior motion (SAM) of anterior mitral leaflet (AML) in M-mode.

dystrophic type in 3 (20%) cases (Table).

### Discussion

DCM was the commonest cardiomyopathy in our series. In a series of 50 children admitted with cardiac failure over a nine-month period, at the Mayo Hospital Lahore, acute myocarditis was the commonest ( $n=24$ ; 48%), while DCM was far behind ( $n=7$ ; 14%).<sup>3</sup> In a retrospective analysis of 100 cases (mean age 54 years) of congestive cardiac failure at the Khyber Teaching Hospital, Peshawar, over an eight-month period, cardiomyopathy was reported in 11 (11%) cases.<sup>4</sup> In an echocardiographic series of 6620 cases of congestive cardiac failure over a 5-year period at the Punjab Institute of Cardiology, Lahore, out of 2335 cases of acquired heart disease, myocardial disease (myocarditis or DCM) was present in 572 (24%) cases.<sup>5</sup>

During a two-year period, 62 children admitted with cardiomyopathy at the Gomal Medical College Hospital, Dera Ismail Khan, DCM was present in 54 (90%), HCM in 4 (7%) and RCM in 2 (3%) cases.<sup>6</sup> In a series of 72 children under 12 years with echo diagnosis of cardiomyopathy

over a two-year period at the National Institute of Cardiovascular Diseases, Karachi, HCM was reported in 31 (43%), endocardial fibroelastosis in 18 (25%), RCM in 10 (14%) and DCM in 7 (10%) cases.<sup>7</sup> In this retrospective study, the higher incidence of HCM and endocardial fibroelastosis could be accounted for by possible filtering-back of the DCM at the referring centres, as being medically treatable not requiring intervention at a tertiary cardiac care centre, and/or overlooked to be myocarditis.

In a study over 9 months in 200 cases of congestive cardiac failure, at the Liaquat University of Medical and Health Sciences, Jamshoro, DCM was reported in 140 (70%) cases and HCM in 22 (11%) cases.<sup>8</sup> In a series of 66 cases of sudden cardiac death (21-58 years) over a 3-year period, autopsied at the Armed Forces Institute of Pathology, Rawalpindi, myocardial hypertrophy (symmetrical and asymmetrical) was confirmed in 5 (7.5%) cases.<sup>9</sup> In a series of 1019 cases of congestive cardiac failure, at the Lady Reading Hospital, Peshawar, over 30-month period, DCM was present in 103 (10%) cases and obstructive and restrictive cardiomyopathy in 51 (5%) cases.<sup>13</sup>

In 50 cases admitted for congestive cardiac failure at the Khyber Teaching Hospital Peshawar, on echocardiography DCM was reported in 27 (54%) cases.<sup>14</sup> Over 18 months in 1166 cases examined for heart disease at the Pakistan Institute of Medical Sciences, Islamabad, out of 229 (19%) cases of acquired heart disease, DCM was present in 39 (17%) cases.<sup>15</sup> Over a two-year period, 62 patients with left bundle branch block-type broad complex tachycardia, seen at the Hayatabad Medical Complex, Peshawar, 12 (19%) cases were confirmed to have ARVC, and the right ventricle was dilated in 11 (92%) cases.<sup>16</sup>

In a study of subjects in the last months of pregnancy, conducted over 4 years at the Civil Hospital, Karachi, PCM was diagnosed in 32 cases, i.e. at the rate of 1 per 837 deliveries.<sup>10</sup> During two years at the Sheikh Zahid Hospital, Larkana, out of 25 cases of PCM, 19 (76%) presented during the post-partum period. On echocardiography, 17 (70%) cases had EF < 30%, and 74% had left ventricular end diastolic diameter (LVEDD) >55mm.<sup>11</sup> Complete recovery was seen in 12 (48%) cases, and 5 (20%) had persistent cardiomyopathy; 8 (32%) patients died. During 19 months at the Lady Willingdon and Aitchison Hospitals, Lahore, in 25 cases with PCM, on echocardiography the mean EF was 20-25% with global hypokinesia, and 10 (40%) cases had mild mitral and tricuspid regurgitation.<sup>12</sup>

In a series of 129 cases of PCM, over a year at the Ayub Medical College Hospital, Abbotabad, on echocardiography 71 (91%) cases had dilated left ventricle with systolic dysfunction.<sup>17</sup> In 30 patients with PCM, at the Liaquat University Medical and Health Sciences, Jamshoro, in 21 (70%) cases the LVEDD was >57 mm in 21 (70%) with reduced EF <40%. Nine (30%) cases had normal sized left ventricle with generalised left ventricular hypokinesia and reduced EF (<40%); 18 (60%) patients had moderate mitral regurgitation.<sup>18</sup> Over a 21-month period at the National Institute of Cardiovascular Disease, Karachi, in 35 patients with PCM, two dimensional (B-mode) and Doppler assessments were carried out at the time of diagnosis and 6 months later for assessment of recovery;<sup>19</sup> 25 (71%) cases had EF <30% and 26 (74%) had LVEDD >55mm; 9 (26%) cases had recurrent PCM.

## Conclusion

DCM was the most frequently diagnosed subtype of cardiomyopathy followed by HCM in both the paediatric and adult age group.

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