FREQUENCY OF LEFT VENTRICULAR NON-COMPACTION IN PATIENTS WITH CONGESTIVE CARDIOMYOPATY

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ABSTRACT

Background: Left ventricular noncompaction (LVNC) is a recently and increasingly recognized form of cardiomyopathy. It is caused by failure of developing myocardium to become compact thereby resulting in "spongy myocardium". The objective of this study was to find the frequency of left ventricular non compaction in patients with congestive cardiomyopathy.

Material & Methods: This cross sectional study conducted in Cardiology Unit, Hayatabad Medical Complex, Peshawar from March 2009 to August 2009. One hundred and fifty patients of congestive cardiomyopathy were included. Relevant history about duration of symptoms and medication was obtained. ECG was recorded to rule out ischemic heart disease. We confirmed congestive cardiomyopathy by echocardiography with left ventricular end diastolic dimension of more than 5.6 cm and ejection fraction of less than 30%. Echo was carried out to determine the left ventricular wall thickness, internal dimension and left ventricular non compaction of congestive cardiomyopathy of other causes were excluded eg; ischemic heart disease, hypertension and valvular heart disease etc..

Results: One hundred and fifty individuals were included in this study. Male patients were 98 (65.3%) and female were 52 (34.7%). Male to female ratio was about 3:1. Mean age was calculated as 54 years with standard deviation as ±11.3. Thirteen (8.6%) patients were in age group between 20-40 years, followed by 101 (67.3%) patients in range of 40-60 years and 36 (24%) patients were between 60-80 years. LV non compaction noted in 3 (2%) patients out of 150 patients.

Conclusion: Left ventricular noncompaction (LVNC) is a form of cardiomyopathy that should be considered as a possible diagnosis because of its potential complications. Echocardiography is the standard tool for diagnosis.

Key Words: Trabeculation, Ventricular non-compaction, Cardiomyopathy.

INTRODUCTION

Isolated non-Compaction of left ventricle is a congenital cardiomyopathy. It is relatively recently recognized entity categorized as "unclassified cardiomyopathy" by World Health Organization[1].It is caused by failure of developing myocardium to become compact thereby resulting in "spongy myocardium"[2].The current literature suggests that LVNC in adults is rare and associated with poor prognosis[3,4]. In children, LVNC can occur in Barth syndrome, a rare X-linked multi-system disorder caused by a mutation in the G4.5 gene that encodes the tafazzin family of proteins[5].However, mutations in this gene in adult LVNC are rare.[6,7]

Three major clinical manifestations of noncompaction have been described: Heart failure, arrhythmias, and embolic events[8,10]. Over two thirds of the patients in the largest series with LVNC had symptomatic heart failure[11]. Arrhythmias are common in patients with ventricular noncompaction. Atrial fibrillation has been reported in over 25% of adults with LVNC.[8,11]. Ventricular tachyarrhythmia has been reported in as many as 47%. Sudden cardiac death accounted for half of the deaths in the larger series of patients with non-compacted LV.[8,10,11,12].The occurrence of thromboembolic events, including cerebrovascular accidents ,transient ischemic attacks, pulmonary embolism, and mesenteric infarction, ranged from 21% to 38%.[8,10,11].

Transthoracic echocardiography and magnetic resonance angiography are the diagnostic methods of choice for non-compaction cardiomyopathy. Ventriculography and computed tomography may also show the typical morphological features of non-compaction: pathognomonic combination of, firstly, multiple prominent ventricular trabeculations and, secondly, multiple deep intertrabecular recesses communicating with the ventricular cavity. Most commonly, apical and midventricular segments of both the inferior and lateral wall

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were affected in more than 80% of patients[13]. Involvement of the midventricular anterior wall and septum and the basal segments is much less frequent.

Treatment for noncompaction of the ventricular myocardium focuses on the 3 major clinical manifestations: Heart failure, arrhythmias,and systemic embolic events. Standard medical therapy for systolic and diastolic ventricular dysfunction is warranted. Cardiac transplantation has been used for those with refractory congestive heart failure[15].The beneficial effects of the ß-blocker on left ventricular function, mass, and neurohormonal dysfunction in an infant with LVNC have been described[16]. Prevention of embolic complication is recommended as long-term prophylactic anticoagulation for all patients with ventricular noncompaction whether or not thrombus has been found.[8,11] Because of the familial association described with noncompaction, screening echocardiography of first degree relatives is recommended.

We carried out this study with the aim was to find out the frequency of left ventricular non compaction in our patients with congestive cardiomyopathy.

MATERIAL AND METHODS

This descriptive cross sectional study was conducted in Cardiology Unit, Hayatabad Medical Complex, Peshawar from March 2009 to August 2009. One hundred and fifty patients of congestive cardiomyopathy were included. We took patient from both outpatient and inpatient. Informed consent was taken by explaining risk and benefit ratio of the study. Relevant history about duration of symptoms and medication was obtained. ECG was recorded to rule out ischemic heart disease. We confirmed congestive cardiomyopathy by echocardiography with left ventricular end diastolic dimension of more than 5.6 cm and ejection fraction of less than 30%.Echo was carried out to determine the left ventricular wall thickness, internal dimension and left ventricular non compaction according to the Chin et al.[21] Confounding variables were controlled by excluding other causes (ischemic heart disease, hypertension, valvular etc.) of congestive cardiomyopathy.

On the 2-D echocardiographic images, trabeculations were defined as localized protrusions of the ventricular wall _>3 mm in thickness associated with intertrabecular recesses filled with blood from the LV cavity (visualized by colour Doppler). When two myocardial layers could be identified, the presence of numerous small cavities within the inner myocardial layer was confirmed using colour flow Doppler.

Three echocardiographic definitions were used for the identification of LVNC in the heart failure and control groups. The size and number of trabeculations were evaluated on the apical views in diastole and the thickness of the non-compacted layer was measured on parasternal short-axis views in systole.

1. Chin et al.21

LVNC is defined by a ratio of X/Y _< 0.5

X = distance from the epicardial surface to the trough of the trabecular recess

Y = distance from the epicardial surface to peak of trabeculation

These criteria focus on trabeculae at the LV apex on the parasternal short axis and apical views, and on left-ventricular free-wall thickness at end-diastole.

2. Jenni et al.22

(i) A two-layer structure, with a thin compacted layer and a thick non-compacted layer measured in end systole at the parasternal short-axis views

LVNC is defined by a ratio of N/C.2 where

N = non-compacted layer of myocardium

C = compacted layer of myocardium

(ii) Absence of co-existing cardiac structural abnormalities

(iii) Numerous, excessively prominent trabeculations and deep intratrabecular recesses

(iv) Recesses supplied by intraventricular blood on colour Doppler

3. Stollberger et al.12

(i) More than three trabeculations protruding from the left-ventricular wall, apically to the papillary muscles, visible in a single image plane

(ii) Intertrabecular spaces perfused from the ventricular cavity, visualized on colour Doppler imaging

RESULTS

One hundred and fifty individuals were included in this study.Thirteen (8.6%) patients were in age group between 20-40 years, followed by 101 (67.3%) patients in age range of 40-60 years and 36 (24%) patients were in age group of 60-80 years. Mean age was calculated as 54 years. (shown in table).

Male patients were 98 (65.3%) and female were 52 (34.7%). Male to female ratio was about 3:1 (as shown in table).

NYHA Class I/II noted in 31(20.7%) patients with LVNC, and 119(79.3%) patients presented with NYHA Class III/IV. (shown in table.)

LV non compaction was observed among 3(2%) patients out of one hundred and fifty.

DISCUSSION

LVNC is defined by the presence of prominent trabeculations on the luminal surface of ventricle in

Table: Characteristics of patients. (N= 150)

	No. of Patients	Percent
Age (years)		
20-40	13	8.6
41-60	101	67.4
61-80	36	24
Gender		
Male	98	65.3
Female	52	34.7
LV Non Compaction		
Yes	3	2
No	147	98
NYHA functional Class		
Class I/II	31	20.7
III/IV	119	79.3

association with deep recesses that extend into the ventricular wall but which do not communicate with the coronary circulation.[17] In some cases, LVNC occurs in association with other congenital heart defects, including atrial and ventricular septal defects, congenital aortic stenosis, and aortic coarctation 18 when no other congenital heart lesion is present, LVNC is said to be isolated. Until recently, isolated LVNC was thought to be extremely rare with a prevalence in adults of <0.3% [17, 19] and an annual incidence in children of <0.1 per 100 000.[18,20] However, the exponential rise in the number of reports of patients with LVNC suggests that an increased awareness and the use of modern ultrasound technology have resulted in an increased detection of the morphological features of LVNC in routine clinical practice.

Diagnostic criteria for left-ventricular non-compaction were proposed through a number of echocardiographic definitions for the diagnosis of LVNC. The method proposed originally by Chin et al.[21], evaluates the size of trabeculations in relation to the thickness of the compacted wall in different echocardiographic views and at different levels of the left ventricle in end-diastole. Jenni and coworkers,[22] have proposed a method that relies on the detection of two myocardial layers, compact and non-compact, in short-axis views in absence of other congenital heart lesion, LVNC is said to be isolated. In this study, we used Chin et al.[21] echocardiographic definitions to identify LVNC, assessing the number and size of trabeculations, as well as the relative thickness of the non-compacted layer when possible. The third definition, proposed by Stöllberger et al determines the number of prominent trabeculations visible in apical views of the left ventricle in diastole. 12In this study, we used Chin et al.[21] echocardiographic definitions to identify LVNC, assessing the number and size of trabeculations, as well as the relative thickness of the non-compacted layer when possible. It has been suggested that the specificity of the echocardiographic criteria for LVNC can be improved by applying additional morphologic parameters such as the presence of an extensive meshwork of trabeculae.[20]

Increasingly, CMR imaging is also being used to detect LVNC. CMR has the advantage of good spatial resolution at the apex and lateral wall of the left ventricle and, in a recent study, has been used to quantify the ratio of non-compacted and compacted layers in patients with LVNC and in normal controls.[24] However, most CMR studies have used an adapted version of existing echo criteria with all the same limitations. Moreover, direct comparison with echo is not always possible as the detection of the non-compact layer using CMR is performed in diastole, whereas the echocardiographic definition applies to systole (in the case of the Jenni criteria). Fine trabeculations are a feature of the normal left ventricle, but it is only quite recently that technical advances such as harmonic imaging, contrast echocardiography and CMR imaging have permitted their detection in normal individuals.

Early reports suggested that LVNC is associated with severe LV dysfunction and a high incidence of ventricular arrhythmias and thrombo-embolic complications.[19] More recent studies describe a much lower incidence of death, stroke, or documented sustained ventricular arrhythmia.[25] Only longitudinal studies can determine the additional thrombo-embolic and arrhythmic risk posed by the presence of prominent trabeculae in dilated ventricles.

CONCLUSION

Left ventricular noncompaction (LVNC) is a recently and increasingly recognized form of cardiomyopathy that should always be considered as a possible diagnosis because of its potential complications. Echocardiography is the standard tool for diagnosis, and CMR is very useful to confirm or rule out this disease, especially when the apex is difficult to visualize. Nevertheless, there are still many questions regarding this disease that require further investigation and follow up.

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