Noncompaction cardiomyopathy in the State of Qatar

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ABSTRACT

Objectives: To study the development of noncompaction of the ventricular myocardium (NCCM) in the state of Qatar and to highlight the prognostic parameters in those patients.

Methods: We conducted this study from 2000 to 2004 on patients who were referred to Hamad General Hospital with questionable echocardiographic features of cardiomyopathy with or without clinical manifestations of heart failure and were found to have NCCM. The diagnosis of NCCM was made according to echocardiographic criteria in 12 cases and those patients are followed up for 2-5 years.

Results: The mean age at diagnosis of NCCM was 6.5 years. Among them, 4 were males and 8 were females. Family history of NCCM was reported in 5 cases. Normal ejection fraction was detected in 5 patients; in this group pulsed-Tissue Doppler Imaging revealed evidence of subclinical systolic dysfunction in 4 cases. All patients showed variable degrees of diastolic dysfunction. Severely impaired ejection fraction was found in 3 cases. Progression to dilated cardiomyopathy occurred in 4 cases. Site of noncompaction included left ventricle apex in all cases, inferoposterior in 11 cases, and lateral wall in 11 cases while biventricular noncompaction was noted in 4 cases. Electrocardiogram findings included right bundle branch blocker (3) patients, left bundle branch blocker (2), left ventricular hypertrophy (6) and right ventricular hypertrophy in 3 cases. Atrial tachyarrhythmias developed in 4 cases. Wolff-Parkinson-White syndrome was detected in one patient. Associated congenital anomalies included ventricular septal defect, pulmonary stenosis, aortic coarctation, and Ebstein anomaly. The overall mortality rate was 25%.

Conclusion: Noncompaction cardiomyopathy is so rare to be easily missed. The prognosis is poor in symptomatic cases; however, detection of subclinical systolic dysfunction is needed.

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Noncompaction of the ventricular myocardium (NCCM) is a rare entity characterized by multiple prominent trabeculations with deep intertrabecular recesses producing the characteristic spongy appearance resulting from arrest of myocardial compaction during fetal life.[1] Noncompaction associated with congenital heart disease has been shown to result from mutations in the alpha-dystrobrevin gene and transcription factor NKX2.5.[2] Diagnosis of NCCM is made with 2-dimensional echocardiography, cardiac magnetic resonance imaging, or LV angiography.[1,2] Although NCCM has been known for more than a decade, it is still an "unclassified" cardiomyopathy according to the World Health Organization classification of the cardiomyopathy.[3] The diagnosis of this disorder is mostly missed. Early diagnosis of NCCM and correct management of such patients are crucial as the clinical manifestation is characterized by important morbidity and mortality caused by early heart failure, life threatening ventricular arrhythmias and systemic embolic events.[4,5] The fact that NCCM is often detected retrospectively in patients who had already been diagnosed with DCM suggests that its frequency in the heart failure population may have been underestimated. This underestimation is probably related to inadequate imaging of the apical segments of the left ventricle (LV) and recent reviews have noted an increased incidence of NCCM with improving cardiac imaging.[6] Lack of physician awareness may have also led to an underestimation of the incidence of LVNC. Of the 2 published criteria for NCCM available, Jenni criteria[7] stress the presence of a 2-layered structure, whereas Chin criteria[1] focus on the depth of recess compared with the height of trabeculae. Murphy et al[8] suggested that isolated NCCM could be classified as a sub-type or variant of idiopathic DCM rather than a distinct cardiomyopathy

in itself. In our study, we focused on the prevalence and clinical presentation of this entity in Qatar. We discuss some of the new insights into risk stratification and the parameters that presage poor prognosis in patients with NCCM.

Methods. This study is conducted from 2000 to 2004 on patients who were referred to Hamad General Hospital with questionable echocardiographic features of cardiomyopathy with or without clinical manifestations of heart failure and were found to have NCCM. The diagnosis of NCCM was made according to echocardiographic criteria in 12 cases and those patients are followed up for 2-5 years. The data includes personal and family history; physical examination; 12-lead electrocardiogram (ECG); chest radiograph; and M-mode, 2-dimensional, and Doppler echocardiographic examinations and tissue Doppler imaging. A diagnosis of noncompaction of ventricular myocardium was made on the basis of the presence of numerous, excessively prominent trabeculations associated with deep intertrabecular spaces by ECG. To quantify the extension of trabecular meshwork, the thickness of ventricular wall and X-to-Y ratios were measured as reported by Chin et al.[1] The left ventricular end diastolic diameter, ejection fraction (EF) and fractional shortening (FS) were measured by M-mode echocardiography. The measurements were carried out in parasternal long-axis view, just beneath the mitral valve and at the level of papillary muscle. The inner wall of the LV was traced as the trough of the recess in systole and diastole. Pulsed tissue Doppler imaging was carried on cases by measuring systolic myocardial(S_m) and early (E_m) and late (A_m) diastolic myocardial velocities in LV and right ventricle (RV) to assess the systolic and diastolic function. A Sm velocity <9.0 mm was considered an indicator of systolic dysfunction.[9] Written informed consent was obtained from all patients.

Results. Since 2000, 12 cases (8 females and 4 males) of LV noncompaction were identified at our institution. They were followed up for 2-5years. The mean age at diagnosis was 6.5 years, 9 patients aged <10 years, 2 patients 10-20 years, one patient aged 37-year-old, and no consanguinity has been recorded. Family history of noncompaction was reported in 5 cases,[1,8,10-12] and history of DCM in one case.[9] Shortness of breath was the most common presentation (58%), and then palpitation (17%). Noncompaction was diagnosed accidentally during check up for relatives

Table 1 - Clinical characteristics of patients with noncompaction cardiomyopathy at presentation.

Clinical characteristics	Patients number											
	1*	2*	3*	11*	12*	4†	7†	9†	10†	5‡	6‡	8‡
Gender	F	F	F	M	F	F	F	M	M	F	F	M
Age (years)	7	4	6	7	15	3	23	4	37	2	3	5
Familial cardiomyopathy	Yes	No	No	Yes	Yes	No	No	Yes	Yes	No	No	Yes
Consanguinity	No	No	No	No	No	No	No	No	No	No	No	No
Presentation	Accidentally	Accidentally	SOB	Palpitation	Accidentally	SOB	SOB	SOB	Palpitation	SOB	SOB	SOB
Congenital anomaly	PS	VSD	VSD	No	No	Ebstien	No	No	No	VSD	VSD, Aortic Coarctation	No
BBB	NL	Rt	Lt	Rt	Rt	No	Lt	no	No	No	No	No
Ventricle hypertrophy	No	No	No	No	Lt	Lt	Lt	Lt+Rt	Lt	No	Rt	Lt+Rt
Ejection fraction	76%	77%	66%	60%	76%	50%	45%	35%	40%	30%	30%	20%
LV apical NC	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Lateral wall NC	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Infroposterior NC	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes
RV involvement	No	No	No	No	Yes	No	No	No	No	Yes	Yes	Yes
Sm lat.wall(cm)	9	6	6.5	8	6.6							
Sm RV(cm)	12	8	13	13	13							
Arrhythmia	No	No	No	No	No	No	No	No	Yes	Yes	Yes	Yes
CHF	No	No	No	No	No	No	No	Yes	No	Yes	Yes	Yes
Outcome	A	A	A	A	A	A	A	A	A	Died	Died	Died

*Normal systolic function, †mild to moderate impaired systolic function, ‡severely impaired systolic function. BBB - bundle branch blocker, Rt - right, Lt - left, LV - left ventricle, A - alive, RV - right ventricle, NC - noncompaction, Sm - myocardial systolic velocity, CHF - congestive heart failure, SOB - shortness of breath, NL - normal

or due to cardiac murmurs in 25% of our cases. Facial dysmorphism has been noted in one case in the form of low set ear and epicanthal folds. Normal EF (>60%) was detected in 5 patients[1-3,11,12] (**Table 1**). For those patients pulsed-Tissue Doppler Imaging (TDI) revealed evidence of subclinical systolic dysfunction in 4 cases (Sm <8 mm). All patients showed variable degrees of diastolic dysfunction but restrictive pattern was noted in 40% of cases and was associated with poor prognosis and early death in one case. Mild to moderate systolic dysfunction (left ventricular ejection fraction 50% to >30%) was diagnosed in 4 cases while severely impaired EF was present in 3 cases (**Table 2**). Ejection fraction was normalized in one patient only. Progression to dilated cardiomyopathy occurred in 4 cases.[6-9] Site of noncompaction was variable: 4 patients had biventricular noncompaction.[5,6,8,12] This carried poor prognosis as 3 out of 4 died. Left ventricle apex was involved in all cases, inferoposterior wall in 11 cases, and lateral wall in 11 cases. **Figure 1** demonstrates different echocardiographic findings of NNCM. There was no evidence of thromboembolic events in our cases. At presentation, ECG was normal in 2 patients; right bundle branch block was detected in 3 patients, left bundle branch block in 2 patients, left ventricular hypertrophy (LVH) in 6 patients and right ventricular hypertrophy (RVH) in 3 patients (**Table 1**). Atrial tachyarrhythmias developed in 4 cases (2 sinus tachycardia and 2 supraventricular tachycardia [SVT]) and this was the most related parameter to poor prognosis (3 patients died). Wolff-Parkinson-White syndrome (WPW) was detected in one patient who was treated successfully by radiofrequency ablation. Associated congenital anomalies included ventricular septal defect (VSD) that was the most common findings (4 patients), pulmonary stenosis (PS) in one patient, aortic coarctaion in one patient, and Ebstien anomaly in one patient. All males had family history of cardiomyopathy, and 50% of them progressed to DCM. Twenty-five percent had biventricular noncompaction and 25% died. Estimated mortality rate is 25% (3 cases: 2 females and one male), all aged <5 years (**Table 1**) after the diagnosis. All patients developed congestive heart failure (CHF) before death (**Table 1**).

Discussion. Qatar is a small peninsula that is characterized with high consanguinity rate. The incidence rate of all types of cardiomyopathies in the State of Qatar is 2.5/100,000 populations per year.[10] In the present study, we recorded 12 cases of NCCM.

Table 2 - Clinical characteristics of patients with noncompaction cardiomyopathy in different studies.

Clinical characteristics	Chin et al[1]	Ritter et al[4]	Ichida et al[11]	Oechslin et al[5]	Stollberger et al[23]	Ozkutlu et al[24]	Murphy et al[8]	Pignatelli et al[25]	Present study
Patient no.	8	17	27	34	62	12	45	36	12
Male (%)	63	82	56	74	70	91	62	55	33
Age range	0.9-22	18-71	0-15	16-71	18-75	0-11	13-83	1d-17ys	0.4-34
Follow up (years)	<5	<6	<17	<11	<6	<10	<10	0.5-12	5
Familial occurrence of cardiomyopathy (%)	50	12	44	18	-	-	51	30	50
Site of noncompaction (%)									
LV: Apical	Prominent	100	100	94	98	N/A	N/A	78 (LV	100
Inferior wall	-	100	70	84	8	N/A	N/A	alone)	91
Lateral wall	-	-	41	100	19	N/A	N/A		91
RV	-	-	-	-	-	N/A	N/A	22 (both ventricle)	33
Bundle branch block (%)	25	47	15	56	26	8	29	N/A	41
WPW (%)	13	0	15	0	3	30	0	17%	8
CHF (%)	63	53	30	68	73	30	66	39%	33
Death (%)	38	47	7	35	-	0	2	13%	25
Thromboembolic (%)	Yes	Yes	No	Yes	-	No	Yes	1patient	No
Arrhythmia (%)	Ventricular 66	Ventricular		Ventricular		SVT 30	Ventricular 20 AF 6	2 SVT,1VT	SVT 17

PS - pulmonary stenosis, VSD - ventricular septal defect, SOB - shortness of breath, NL - normal, RBBB - right bundle branch block, LBBB - left bundle branch block, LV - left ventricle, RV - right ventricle, LVH - left ventricular hypertrophy, RAE - right atrial enlargement, LAE - left atrial enlargement, RAD - right axis deviation, DCM - dilated cardiomyopathy, CHF - congestive heart failure, RVH - right ventricular hypertrophy, SVT - supraventricular tachycardia, WPW - Wolff-Parkinson-White syndrome, NCCM - noncompaction cardiomyopathy, AF - atrial fibrillation

Table 3 - Parameters with unfavorable prognosis in noncompaction of the ventricular myocardium.

Parameters
Positive family history of cardiomyopathy
Heart failure at presentation
Restrictive diastolic dysfunction
Sinus tachycardia
Atrial standstill[26]
Persistent atrial fibrillation
New onset left bundle branch block during follow-up
Ventricular tachycardia
Biventricular noncompaction

The true prevalence of NCCM is unclear, and differs from 0.05-0.24% per year but because this represents a population referred for abnormal echocardiographic findings or CHF, it results in selection bias.[4-6] Men appear to be affected more often than women, with males accounting for 56-82% of cases in the 4 largest reported series of NCCM.[1,4,5,11] This was not consistent with our study in which the majority of NCCM cases occurred in females (67%). **Table 2** shows the clinical characteristics of patients with noncompaction cardiomyopathy in different studies. In this rare congenital anomaly, the LV is uniformly affected, but RV noncompaction was described in less than one-half of patients.[4,12,13] The LV apical and inferior wall segments were involved in all patients in an adult population with NCCM studied by echocardiography while the RV apex was involved in 41%.[4] Biventricular noncompaction was shown in 33% of our cases. Noncompaction of the ventricular myocardium can be easily diagnosed by echocardiography with excellent agreement with the necropsy findings if the echo cardiographer is familiar with this disorder and if clear-cut diagnostic criteria are used.[4-5] However, prominent LV trabeculations can be found in up to 68% of healthy hearts and can be observed in hypertrophic hearts secondary to dilated, valvular, or hypertensive cardiomyopathy.[5,14] In a nationwide survey in Japan, Ichida et al[11] reported that the diagnosis of NCCM was missed in 89% of children. Ritter et al[4] observed a mean time from onset of symptoms to correct diagnosis of >3 years in one adult population with NCCM. In our study, 6 patients had congenital heart defects associated with NCCM. Among those, Ebstein anomaly was recorded in one case, which is the fourth reported case associated with NCCM worldwide.[15] Arrhythmias are common in patients with NCCM such as AF (25%), ventricular tachyarrhythmias (47%) while sudden cardiac death accounted for half of the deaths in the larger series of

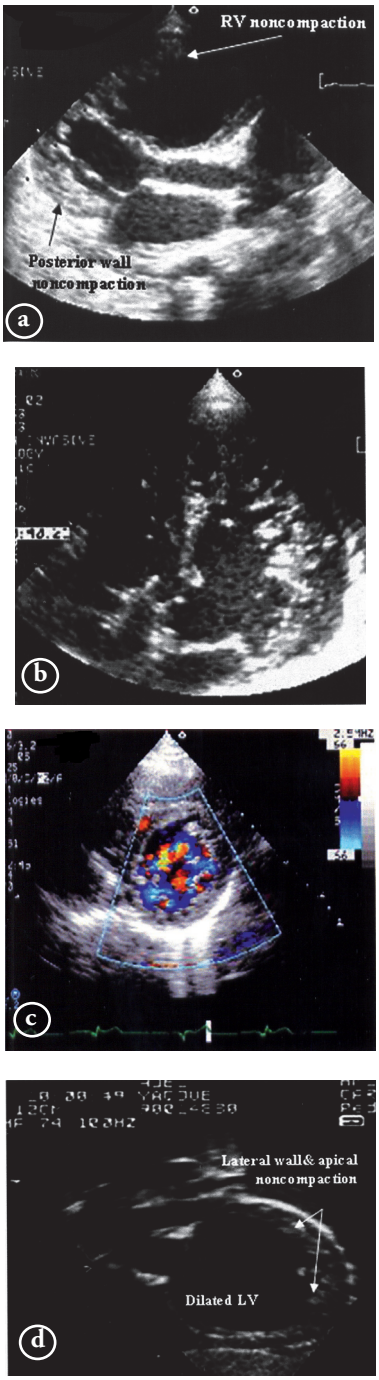

Figure 1 - Demonstrates the sites of noncompaction by 2-echocardiography in the present study in different patients: **a)** parasternal long axis shows LV posterior wall and RV noncompaction **b)** apical view shows the involvement of lateral wall and apex of LV **c)** The parasternal short-axis view of the left ventricle (LV) is shown, the myocardium is thickened, with prominent trabeculations and deep intertrabecular recesses at the level of the papillary muscles. Color Doppler showed flow between the LV cavity and the recesses. **d)** Subcostal transverse view shows dilated LV and lateral and apical noncompaction.

patients with NCCM.[1,4,5,16] Ichida et al[11] did not report ventricular tachyarrhythmias or SCD. Paroxysmal SVT and complete heart block have also been reported in patients with NCCM.[4,11] We report SVT in 2 cases and sinus tachycardia in 2 cases; the latter was the most relevant parameter to poor prognosis. Electrocardiographic findings of the WPW syndrome have been described in up to 15% of pediatric patients,[11,17] but it was not observed in the 2 largest series of adults with isolated NCCM.[4,5] We detected only one patient with WPW. One of our cases presented with atrial standstill. Left bundle branch block has been described in 44% of adult patients with NCCM,[5] but the reported incidence in children was much lower in another study.[11] In our study, LBBB was detected in 2 patients. In the largest multicentered study from Japan[11] pediatric LBBB and ventricular tachycardia were reported rare, more so than that described in adults. The development of LBBB during the follow-up of NCCM, who initially presented with normal ECG, was correlated with the late occurrence of progressive endocardial fibroelastosis in one study.[18] In 3 groups of patients with isolated NCCM, the occurrence of thromboembolic events, ranged from 21-38%.[1,4,5] Embolic complications may be related to the development of thrombi in the extensively trabeculated ventricle, with depressed systolic function, or due to the development of AF.[4,19] Neither our study nor the largest pediatric series with NCCM reported systemic embolic events.[11] Both familial and sporadic forms of NCCM have been described.[1,5,11,20] In the largest reported sereies, familial recurrence was seen in 18%.[5] This may be underestimated because of incomplete screening of siblings. It approaches 50% in our study. In patients with familial disease, relatives may have features consistent with dilated cardiomyopathy rather than NCCM.[8] Clinical presentation varies from asymptomatic LV dysfunction to severe, disabling CHF. Over two-thirds of the patients in the largest series with NCCM had symptomatic heart failure,[5] 33% of our patients developed CHF. Left ventricular dysfunction may develop, regardless of the presence or absence of symptoms at initial diagnosis.[11] The origin of systolic dysfunction in NCCM may relate to subendocardial hypoperfusion and microcirculatory dysfunction even in the absence of epicardial coronary artery disease.[1] Diastolic dysfunction in NCCM may be related to both abnormal relaxation and restrictive filling caused by the numerous prominent trabeculae.[19] Initial presentation of NCCM as a restrictive cardiomyopathy has been described in children with NCCM.[11,21] All our patients showed variable degrees of diastolic dysfunction but restrictive pattern was associated with the worst prognosis and early death. The role of TDI in NCCM is still not investigated well. Williams et al[22] demonstrated the value of strain and strain rate in the detection of NCCM in a case report. We used the myocardial systolic velocity in those with normal EF to detect subclinical systolic dysfunction. Low myocardial systolic velocities in LV and RV were detected in 80% of cases. There is no definite prognostic criterion for NCCM. **Table 3** summarizes criteria of unfavorable outcome in NCCM. Prognosis for patients with NCCM is variable and nearly 60% of patients described in one large series[4] had either died or undergone cardiac transplantation within 6 years of diagnosis. Murphy et al[8] reported that NCCM is associated with a better prognosis than previously reported. According to Oechslin et al,[5] 53% of patients had heart failure requiring hospitalization, 41% had ventricular tachyarrhythmias, with 12% receiving an implantable cardioverter-defibrillator (ICD), 35% died, and 12% underwent heart transplantation. Patients with high-risk features are candidates for early and aggressive interventions. Medical management varies with the clinical manifestations; left ventricular EF and the presence or absence of arrhythmias. Patients with a reduced EF should be treated with standard medical therapy for this disorder. Anticoagulation therapy is usually recommended in patients with AF and/or EF <40%. Holter monitoring should be considered once a year to detect asymptomatic arrhythmias. The indication for ICD therapy is similar to those in dilated cardiomyopathy. Heart transplantation should be considered for patients with end-stage heart failure,[5, 27]

In conclusion, noncompaction cardiomyopathy is so rare to be easily missed. The prognosis is poor in symptomatic cases. Recognition of subclinical systolic dysfunction is needed to improve the outcome.

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