Epidemiology of Idiopathic Cardiomyopathy in Qatar during 1996–2003

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Key Words

Epidemiology · Consanguinity · Cardiomyopathy · Qatar

Abstract

Objectives: To report the rates of cardiomyopathies in the population below 50 years of age in Qatar. **Subjects and Methods:** We conducted a retrospective review of clinical data on patients with cardiomyopathy who were hospitalized in Hamad General Hospital, Doha. Data were collected from medical records during the 1996–2002 period and prospectively from the patients who were hospitalized during the year 2003. All cardiomyopathy patients below 50 years of age who were citizens or permanent residents in Qatar were included in this study. **Results:** During the study period, a total of 132 cases were recorded with idiopathic cardiomyopathies. Among these, 67.4% were males and 32.6% females; Qatari 31.8%, non-Qatari 68.2%. The consanguinity rate was high among Qatari patients. In the first 7-year study period, 1996–2002, the incidence rate of all types of cardiomyopathies was 2.5/100,000 population per year (95% CI: 1.4–3.5). It increased to 5.2/100,000

population during the year 2003 (95% CI: 3.6–6.7). Dilated cardiomyopathy was most prevalent (75.8%) in all age groups, and the incidence increased remarkably with age. Lower prevalence of hypertrophic cardiomyopathy (13.6%) and left ventricle noncompaction cardiomyopathy (6.1%) was found. In children below 15 years of age, the incidence rate for all types of cardiomyopathies was 2.7/100,000 population. The overall mortality rate was 5.3%. **Conclusion:** Most cases of cardiomyopathy were identified at an early age: below 15 years and above 35 years of age. Introducing preventive and early diagnosis programs may have an impact on reducing the mortality and morbidity from idiopathic cardiomyopathy.

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Introduction

Congestive heart failure (CHF) has been termed the ‘new epidemic of cardiovascular disease’ in the new millennium. In spite of the importance of this disease worldwide, little information is available about the epidemiology of CHF in the developing world [1, 2]. There are no

population-based studies, examining the prevalence and causes of CHF in young patients in Qatar. This information is needed in order to develop strategic planning for prevention and treatment of this rising epidemic. The prevalence of the various types of cardiomyopathies may be different from those reported elsewhere and this may warrant specific strategic planning [3, 4]. Furthermore, the Qatari population, like other Arabian Gulf countries, has consanguineous family structure that is different from most of the world population.

In Qatar, the consanguinity rate is very high. Therefore, the population is in principle genetically homogeneous [5]. This will serve as an ideal population for studying the genetics of cardiomyopathies with respect to the nature of the genetic polymorphism that can influence the power and interpretation of epidemiological data when dealing with more out-bred and heterogeneous populations. This study examined the rates of cardiomyopathies in the population below 50 years of age in Qatar, in the 1996–2003 period.

Subjects and Methods

We analyzed the data with respect to all patients below 50 years of age who were hospitalized with idiopathic cardiomyopathy during the 1996–2003 period. This study was based at Hamad General Hospital, Doha, which is the main tertiary care center in the country. Hence it was an ideal site for population-based studies. All patients with various types of cardiomyopathy were treated at this hospital. In the present study, the subjects were recruited from the Department Cardiology. The data were collected retrospectively from the medical records of the patients during the 1996–2002 period when they visited outpatient clinics for follow-up, and for the year 2003 it were prospective data from the clinical records at the time of patient's hospital discharge. Cases of cardiomyopathy were classified according to World Health Organization/International Society and Federation of Cardiology task force on Cardiomyopathies [6, 7]. The inclusion criteria were subjects aged below 50 years and all nationalities in Qatar presenting with any type of cardiomyopathy. Those suffering from end-stage rheumatic valvular heart disease and CHF complicating acute myocardial infarction were excluded.

The studied subjects had complications of shortness of breath, arrhythmias, easy fatigability, chest pain, thromboembolic manifestations or congestive gastroenteropathy symptoms and echocardiographic features of cardiomyopathy. The diagnosis of dilated cardiomyopathy (DCM) was essentially one of exclusion and was based on clinical symptoms, electrocardiographic and chest roentgenographic findings of ventricular dysfunction, supported by echocardiographic or angiographic confirmation of global ventricular dilatation: left ventricular end diastolic volume more than or equal to 110 cm³/m², left ventricular end systolic volume more than or equal to 44 cm³/m², and left ventricular ejection fraction more than or equal to 50%. The diagnosis of

hypertrophic cardiomyopathy (HCM) was based on specific echocardiographic criteria [8]. Echocardiographic measurements for HCM included left ventricular septal wall thickness, typically >15 mm, dilated left atrium (>4 cm) and left ventricular end-diastolic diameter <45 mm. Normally, for the athletic heart, the septum is <15 mm, the left atrium is <4 cm and left ventricular end-diastolic diameter >45 mm.

Restrictive cardiomyopathy (RCM), in which endomyocardial scarring may affect one or both ventricles restricting ventricular filling was based on echocardiographic findings, and/or the hemodynamic changes during ventriculography are diagnostic [9]. Peripartum cardiomyopathy was defined as CHF that occurs late in pregnancy or during the early postpartum period (the last trimester or up to 3 months postpartum) in the absence of congenital, coronary, or valvular heart disease or another recognized cause of heart failure. Patients were described to have unclassified cardiomyopathy when they did not fit readily into any group defined above.

Diagnosis of isolated ventricular noncompaction was based on the strict criteria of echocardiography. The criteria were an absence of significant heart lesions; segmental thickening of the left ventricular myocardial wall consisting of two layers consisting of a thin, compacted epicardial and an extremely thickened endocardial layer with prominent trabeculations and deep recesses [10].

We reviewed the clinical data of patients hospitalized with cardiomyopathy which included sociodemographic characteristics, symptoms, smoking habits, history of maternal gestational DM (DCM), medication, clinical examination, laboratory tests, chest X-ray, electrocardiogram, echocardiogram, stress test and/or cardiac catheterization (if available), as well as cardiac and/or skeletal biopsy (if needed).

Data are expressed as mean and standard deviation unless otherwise stated. Student's *t* test was used to ascertain the significance of differences between mean values of two continuous variables. Fisher exact test and χ^2 tests were performed to test for differences in proportions of categorical variables between two or more groups. The level *p* < 0.05 was taken as the cut-off value for significance.

Results

During the 8-year study period, 132 patients were diagnosed with idiopathic cardiomyopathy. Of these, 31.8% were Qataris and 68.2% non-Qataris. Of the Qatari patients, 33.3% married first degree relatives and 23.8% second degree relatives, giving a high level of consanguineous marriages.

The sociodemographic features and clinical characteristics of all types of cardiomyopathy by nationality during the study period are shown in table 1. The proportion of males (67.4%) was double that of females (32.6%). A majority of the adult cases were above 35 years of age. On average, in children below 15 years of age, the incidence rate of all types of cardiomyopathies was 2.9/100,000 population. For all types of cardiomy-

Table 1. Sociodemographic features and clinical characteristics of all types of cardiomyopathy in Qatar by nationality

Variables	Qatari (n = 42)	Non-Qatari (n = 90)
Gender		
Male	22 (52.4)	67 (74.4)
Female	20 (47.6)	23 (25.6)
Age group		
≤ 15 years	10 (23.8)	25 (27.8)
16–35 years	9 (21.4)	12 (13.3)
>35 years	23 (54.8)	53 (58.9)
Body Mass Index		
<25 (normal)	14 (34.1)	42 (53.2)
≥ 25 (overweight)	27 (65.9)	37 (46.8)
Smoking	9 (21.4)	27 (30.0)
Cardiomyopathy		
DCM	30 (71.4)	70 (77.8)
HCM	6 (14.3)	12 (13.3)
LVNC	1 (2.4)	7 (7.8)
ARVC	1 (2.4)	0 (0.0)
RVC	1 (2.4)	0 (0.0)
Unclassified cardiomyopathy	3 (7.1)	1 (1.1)
Associations		
Diabetes mellitus	12 (28.6)	3 (3.3)
Hypertension	14 (33.3)	18 (20.0)
Ejection fraction	38.4 ± 15.9	37.9 ± 13.9
Presentation		
Shortness of breath	25 (59.5)	51 (56.7)
Accidentally	8 (19.0)	28 (31.1)
Palpitation	12 (28.6)	12 (13.3)
Easy fatigue	7 (16.7)	10 (11.1)
Chest pain	5 (11.9)	15 (16.7)
Abdominal pain	2 (4.8)	4 (4.4)
Stroke	0 (0.0)	3 (3.3)
Deaths	6 (14.2)	1 (1.1)

DCM = Dilated cardiomyopathy; HCM = hypertrophic cardiomyopathy; LVNC = left ventricle noncompaction cardiomyopathy; ARVC = arrhythmogenic right ventricular cardiomyopathy; RVC = restrictive cardiomyopathy. Figures in parentheses indicate percentages.

opathies, DCM rate was the highest (75.8%), followed by HCM (13.6%) and left ventricle noncompaction cardiomyopathy (6.1%). RCM and ARVC (arrhythmogenic right ventricular cardiomyopathy) were the least (0.8%). The most common presentations for cardiomyopathies were shortness of breath (57.6%), palpitations (18.2%), chest pain (15.2%) and easy fatigability (12.9%). Abdominal pain was the main symptom in 4.6% of the cases, while stroke was an early manifestation in 2.3% of cases. Twenty-seven percent of the patients were smokers.

All types of cardiomyopathies by age group are presented in table 2. DCM was the most prevalent cardiomyopathy in all age groups and the incidence increased remarkably with age. Hypertrophic cardiomyopathy had two peaks; one below 15 years of age and the other in the 36–50 years age group. Nearly half (48.6%) of the children had DCM, 28.6% HCM and 20% left ventricular noncompaction cardiomyopathy.

From 1996 to 2002, the rate of all types of cardiomyopathies remained at 2.5/100,000 population per year (95% CI: 1.4–3.5), but it increased to 5.2/100,000 population in 2003 (95% CI: 3.6–6.7). Also, the prevalence rate of DCM was 17.5/100,000 population, which was much higher than HCM (3.1/100,000 population). Prevalence rates were greater for men, with a male:female ratio of 2:1 for both DCM and HCM. The percentage of deaths due to cardiomyopathy was 5.3%, and all these were in the 16–35 years age group (table 2). Four deaths occurred in cases with DCM and one in RCM. DCM due to thalassemia was associated with the shortest period between diagnosis and death (2 years).

A rare association between cardiomyopathies and certain diseases is shown in table 3. Among those cases, antiphospholipid syndrome and scleroderma have not been reported before.

Table 2. Type of cardiomyopathies by age group during the 1996–2003 period

Age group	DCM	HCM	LVNC	ARVC	RCM	UCM	Total
1–15 years	17	10	7	0	0	1	35
16–35 years	16	2	1	1	1	0	21
36–50 years	67	6	0	0	0	3	76
Total (%)	100 (75.8)	18 (13.6)	8 (6.1)	1 (0.8)	1 (0.5)	4 (3.0)	132 (100)

Table 3. A rare association between cardiomyopathies and certain diseases during the 1996–2003 period in Qatar

Disease	Type of cardiomyopathy	Age years	Sex	EF	Comment	Incidence
Antiphospholipid syndrome	DCM	38	m	21%	normal coronaries	1 (0.8%)
Cron's disease	HCM	1	m	73%		1 (0.8%)
Familial conus dystrophy	DCM	19	m	38%	completely normalized EF in 2 months	1 (0.8%)
Ebstein disease	LV noncompaction	1	f	50%		1 (0.8%)
Hb H disease	DCM	50	f	30%	HCV	1 (0.8%)
Thyroid disorders	DCM	43–47	m/f	30–45%		3 (2.2%)
Myofibrillar myopathy	RCM	16	m	32%	died within 5 years	1 (0.8%)
Thalassemia	DCM	20–21	m	14–21%	died within 2 years	2 (1.5%)
Scleroderma	DCM	39	f	23%	ICD implanted	1 (0.8%)
Acute myeloid leukemia	HCM	10	f	77%		1 (0.8%)

Discussion

There is no previous report describing the epidemiology of idiopathic cardiomyopathy in Qatar. In the 10-year period between 1991 and 2001, 2,342 patients were hospitalized with CHF. Of these, 53% were Qatari with CHF and hypertension [1]. The high consanguinity rate (46%) in Qatar made us register the incidence of idiopathic cardiomyopathy. We had previously reported striking patterns of cardiomyopathy in two Qatari families [1, 2, 4].

Depending on the diagnostic criteria, the reported annual incidence of idiopathic DCM varied between 5 and 8 cases per 100,000 population [4, 11–13]. In our study, the overall prevalence of all types of cardiomyopathy was 2.5/100,000 population per year during the 1996–2002 period, and it doubled to 5.2/100,000 population in the year 2003. This could be explained in part by our greater awareness of such issue and also because, for this year, it was a prospective study. During the study period between 1996 and 2003, the prevalence of DCM and HCM were 17.5 and 3.1 per 100,000 population, respectively. However, the true incidence is probably underestimated, since many asymptomatic cases remain undetected. The age-adjusted prevalence of idiopathic DCM in the United States averages to 36 cases per 100,000 population [14]. In Japan, a nationwide epidemiological survey to estimate the total number of patients with idiopathic cardiomyopathy and the prevalence of the disorder revealed that crude prevalence per 100,000 population was estimated to be 14.0 for DCM, 17.3 for HCM and 0.2 for RVM [15]. The true natural history of idiopathic DCM is difficult to determine, since asymptomatic cardiomegaly may be present for months or years before being de-

tected. Survival data from tertiary referral centers are similar, with a mortality rate ranging between 25 and 30% in 1 year and approximately 50% in 5 years [16–18]. The overall mortality rate in our study was 5.3%.

Most of the patients with cardiomyopathy are usually first seen between the ages of 20 and 50 years, but the disorder may also affect the children and the elderly [17, 18]. The most common initial manifestation is heart failure, which occurs in 75–85% of patients. In most cases initial symptoms include dyspnea on exertion (86%), palpitations (30%) and peripheral edema (29%). Asymptomatic cardiomegaly is detected in 4–13% of patients. Abdominal pain is not an uncommon presentation either. Chest pain on exertion occurs as an initial symptom in 8–20% of patients [18–20], and eventually occurs in approximately 35%. Systemic and pulmonary emboli are the initial manifestations in 1.5–4% of cases. Although atrial fibrillation is more common among patients with embolic events, it is present in fewer than 25% of such patients. Ventricular arrhythmias are common, but syncope and sudden death are rarely the initial presentation of the condition [15–18]. In our study, the more frequent symptoms of cardiomyopathy were SOB (57.6%), palpitation (18.2%), chest pain (15.2%) and easy fatigue (12.9%).

Ideally, all patients with DCM should be shown to be free of coronary artery disease (CAD); however, in practice, coronary angiography is not routinely performed in all cases with CHF. Thus, retrospective studies cannot control the conditions under which patients are recruited or investigated. It follows that most epidemiologic studies of DCM have compromised on the completeness of data on CAD in their subjects. In an Icelandic study, 75% of

Table 4. Reported studies for idiopathic cardiomyopathy

Location	Time period	Type of study	Cases	Rate	Comment
<i>DCM</i>					
Minnesota, USA [14]	1975–1984	Incidence	45	6.0/100,000	Population-based data, age range 24–88 years
Malmo, Sweden [26]	1970–1979	Incidence	89/188*	3.6–7.5*/100,000	56* cases first established at autopsy + 43* cases with probable DCM
W. Denmark [12]	1980–1981	Incidence	41	0.7/100,000	Cases from three cardiology referral centers and pathology department
England [13]	1983–1984	Prevalence	76	8.3/100,000	Questionnaire survey of family practitioners
Qatar	1996–2003	Incidence	100	17.5/100,000	Population-based data with age started from <1 year to <50 years
Australia [24]	1987–1996	Incidence	184	0.73/100,000	Population-based study only in children <10 years of age
<i>HCM</i>					
Minnesota, USA [14]	1975–1984	Incidence	19	2.5/100,000	Population-based data, age range 16–94 years
W. Denmark [12]	1980–1981	Incidence	20	0.4/100,000	Cases from three cardiology referral centers and pathology department
Iceland [21]	1966–1977	Prevalence	11	33/100,000	Extrapolation from cases identified at autopsy
Japan [15]		Prevalence	22	170/100,000	Echocardiographic screening of industrial workers
Qatar	1996–2003	Incidence	18	3.1/100,000	Population-based data with age started from <1 year to <50 years
Australia [24]	1987–1996	Incidence	80	0.3/100,000	Population-based study only in children <10 years of age
<i>LV noncompaction</i>					
Australia [24]	1987–1996	Incidence	29	9.2 % of all cases (314 cases)	Population-based study only in children <10 years of age
Turkey [27]	1991–2001	Prospective	12	6/10,000	A tertiary care centre
Qatar	1996–2003	Incidence	8	6.1% of all cases but, 20% of CM in age <15 years	

patients were shown to have normal coronary circulation [21]. In studies at Mayo Clinic, the incidence of normal coronary circulation for DCM was 58 and 47%, respectively [14]. In our study, we excluded cardiomyopathies complicating myocardial infarction, and only 7 cases were documented to have CAD, confirmed by angiography.

The true incidence of pediatric cardiomyopathy is also unclear. In comparison with the annual incidence of cardiomyopathy in children in a North American (1.3/100,000), a Finnish (0.74/100,000) and an Australian study (1.24/100,000) [21–26], our study showed a higher rate (2.6/100,000) for the 8-year period. This higher incidence in our study might be related to the high consanguinity rate in Qatar (46%). Left ventricle non-

compaction cardiomyopathy constituted 6.1% of all cardiomyopathies and 20% of pediatric and 0.97% of adult cardiomyopathies in Qatar. Worldwide, the incidence of left ventricular noncompaction cardiomyopathy is 0.05–0.24% in adults [25, 27]. Table 4 summarizes some of the reported studies for cardiomyopathy.

Study Limitations

The retrospective part of the study has some limitations. Coronary angiography was not available for a majority of the cases. The diagnosis of RCM was based only on echocardiographic findings. However, cardiac catheterization is essential to confirm the diagnosis of RCM. Genetic analysis was not performed.

Conclusion

This study revealed that the incidence of idiopathic cardiomyopathy was high in children below 15 years and in adults above 35 years. Introducing preventive and early diagnostic programs may have an impact on reducing

the mortality and morbidity rates of idiopathic cardiomyopathy. Routine baseline echocardiographic study and genetic analysis are recommended in families with consanguineous marriages and a history of idiopathic cardiomyopathy.

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