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Cardiac and Neuromuscular Issues of Cardiomyopathies in a Highly Consanguineous Population

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With interest, we read the article by El-Menyar et al. [1] about the epidemiology of idiopathic cardiomyopathy in Qatar. Between 1996 and 2002, they recorded 132 cases with a prevalence of 76% dilated, 14% hypertrophic cardiomyopathy and 6% left ventricular noncompaction/hypertrabeculation (LVHT). Their findings, however, raise several questions and concerns.

It would be interesting to know why only patients below 50 years were included, how the authors differentiated between 'Qatari' and 'non-Qatari' patients and what countries the non-Qataris come from. It is surprising that among Qataris, the gender distribution was equal, whereas among non-Qataris, there was a male preponderance, and that the prevalence of diabetes mellitus, hypertension and LVHT differed between Qataris and non-Qataris. Moreover, the type of cardiomyopathy in the patients who had suffered a stroke should be clarified.

The diagnosis of LVHT was established according to diagnostic criteria published only in the year 2000. How was LVHT diagnosed in patients investigated between 1996 and 2000? Were all recordings of the echocardiographic examinations reviewed or was LVHT only diagnosed after 2000?

The definition of hypertrophic cardiomyopathy is unusual since it includes left atrial enlargement which is not a common diagnostic prerequisite for hypertrophic cardiomyopathy. It would be interesting to know the reason for this modification, and according to which echocardiographic definition restrictive cardiomyopathy was diagnosed.

It is known that the prevalence of LVHT is dependent on the imaging quality of the echocardiographic machine and on the echocardiographer's awareness of the disorder [2]. In this regard, it should be mentioned if the type of the echocardiographic machine changed during the study period and how many different investigators performed the investigations. The finding that LVHT was diagnosed in 20% of the patients below 16 years but only in 1% of the patients above 16 years should be interpreted.

Regarding the follow-up data, it remains unclear, whether all patients were followed. The causes of death should also be mentioned, and an explanation should be given why death occurred more often in Qataris than in non-Qataris.

Cardiomyopathies and LVHT may represent the first clinical manifestations of a skeletal muscle disorder [3]. Thus, it is of interest, if all patients were systematically investigated by a neurologist and how many patients underwent electromyography and muscle biopsy. The results of the neurologic investigation and of the muscle biopsy would also be of interest. Is the patient listed in table 3 (see El-Menyar et al. [4]) with myofibrillar myopathy and restrictive cardiomyopathy identical with a previously published case or is it a new case?

The present study found a higher incidence of cardiomyopathy in children than previous studies in other countries. The authors relate this higher incidence to the high consanguinity rate in Qatar. We have difficulties in following this interpretation because: (a) in their study, there were more non-Qataris (28%) than Qataris (24%) in the age group below 16 years, and (b) data about the rate of consanguinity in the non-Qataris were not given.

In Qatari patients with cardiomyopathy, the rate of consanguinity was 57% and thus higher than the 46% in the general Qatari population. If the authors suspect a genetic background of the cardiomyopathies, it would be interesting to know if family studies were carried out in the patients with cardiomyopathy.

Answering these questions would be helpful for clarifying the etiology of cardiomyopathies.

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Reply

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Cardiomyopathy was most common in young people and they are the most active group in the community. Hence, we recruited subjects below 50 years. Qataris are the nationals and non-Qataris are foreigners who come to Qatar for employment, most of them being mainly from Asian countries: India, Pakistan, Jordan, Palestine and other Arabian Gulf countries. The non-Qatari male population is high because the majority of them do not have their families in Qatar.

The incidence of diabetes mellitus (25%) and hypertension (15%) is high in the State of Qatar [2]. Qataris who are employed in certain jobs such as military and police services and athletes are screened for left ventricular hypertrophy and hypertrabeculation (LVHT). Dilated cardiomyopathy was present in the patients who had suffered a stroke [1]. We reviewed the records of the echocardiographic examinations retrospectively during the period from 1996 to 2002 and the same criteria were used to collect the data prospectively during the period from 2002 to 2003.

Left atrial enlargement did not affect the inclusion criteria of the study. The authors used the echocardiographic definition for restrictive cardiomyopathy suggested by Ammash et al. [3]. The same type of echocardiographic machine (HP) was used. In patients with LVHT, it is difficult to interpret the study finding that 20% of the patients were below 16 years and 1% above 16 years.

There was a follow-up for all patients. Dilated cardiomyopathy was the most common cause of death. Qatari children outnumbered the non-Qatari children because most of the non-Qataris are residing in the State of Qatar without their family. Since cardiomyopathy was most common in young people and the young population in Qatar comprises more Qataris than non-Qataris, deaths occurred more often among Qataris.

Only 1 patient underwent muscle biopsy. All the patients listed in table 3 (see El-Menyar et al. [4]) with myofibrillar myopathy and restrictive cardiomyopathy are published cases.

The higher incidence of pediatric cardiomyopathy was due to the high consanguinity rate in Qatar [5], not only among Qataris, but also among non-Qataris.

It is true that consanguinity was found in 57.1% of the Qataris with cardiomyopathy [2, 6, 7]. In general, 46% of the population is consanguineous. This could be due to the genetic background of cardiomyopathies. No family study was conducted in patients with cardiomyopathy. We are planning to perform research on cardiomyopathy using molecular genetics and linkage analysis in two or possibly three generations in the near future.

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