Bahrain Medical Bulletin, Vol. 32, No. 2, June 2010

Beta Thalassemia Frequency in Bahrain: A Ten Year Study

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Background: Sickle-cell disease and Thalassanemia syndromes impose a significant economic burden on many countries. Their chronic nature makes them one of the leading causes of morbidity and mortality in those countries[1].

Objective: The aim of this study is to estimate the frequency of beta thalassemia among the students in Bahrain from 1999 to 2008.

Setting: Bahraini Secondary Schools, genetic department and laboratory at Salmaniya medical complex.

Design: Prospective study.

Method: The students in the 11th grade (2nd Secondary class) were screened. Data were collected during the annual student screening program. Informed consents were obtained from the parents.

The blood samples were collected for hemoglobin electrophoresis using HPLC instrument.

Result: Sixty thousand students were screened from 1999 to 2008. The mean prevalence of beta thalassemia trait and major were 2097 (3.5%) and 19 (0.032%) respectively.

Conclusion: The frequency of beta thalassemia in Bahrain was found to be low to moderate compared with other Gulf countries such as UAE, Qatar and Kuwait. Sickle cell disease (SCD) is more common than beta thalassemia in Bahrain. Preventive measures remain the best ways for lowering the incidence of these diseases.

Bahrain Med Bull 2010; 32(2):

Genetic diseases such as beta thalassemia are chronic in nature and require costly lifelong care and management strategies. They cause significant health care and psychosocial burdens on the patient, the family, the health care system and the community[1-9].

The frequency of this disease is high in Lebanon, Jordon, Iraq, Palestine, Egypt and other Arab countries. The carrier rate of beta thalassemia of 3.6-4%, 6.24%, and 8% were observed in Oman, Yemen and UAE respectively. Nadkarni et al 1991, Al-Arrayed and Haites 1995 have observed that beta thalassemia carrier rate of 2-4% in Bahrain population[10-23].

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The beta thalassemia is characterized by a reduced production of β-globin chain (β+) or absent production of β-globin chain (β°). This results in an imbalanced α/non-α globin chain production. The molecular diagnosis is essential in these diseases as the inheritance of beta thalassemia might be masked by coinheritance of sickle cell gene and/or alpha Thalassemia.

Thalassemia major patient shows severe anemia in the first year of life, and are unable to maintain the hemoglobin level of 5 gm/dl. Thus, they need life long blood transfusion, which causes iron overload. Hence, iron chelating treatment is necessary to prevent iron overload damage to the internal organs. In general, the disease may affect the spleen (often enlarged), and causes heart failure.

In recent years, bone marrow transplant and stem cell transplant have shown success in some patients of thalassemia major. Successful transplant can eliminate the patient's dependencies on transfusions.

Most of the beta thalassemia heterozygote carriers are clinically asymptomatic with distinctive hematological phenotype represented by hypochromic, microcytic anemia and characteristically raised levels of HbA2[8-9].

Falciparum malaria was endemic in Bahrain until 1970, and soon afterward eradication was successful. The malaria associated genetic defects of red cells, such as SCD, Thalassaemia and glucose 6-phosphate dehydrogenase deficiency (G6PD) were expected to be common.

The aim of this study is to estimate the frequency of beta thalassemia among the students in Bahrain.

METHOD

The students in the 11th grade (2nd Secondary class) were screened. Data were collected during the annual student screening program. Informed consents were obtained from the parents.

The blood samples were collected for hemoglobin electrophoresis using HPLC instrument**.** G6PD deficiency was also tested.

The plan was to screen all the students in the 11th grade (2nd Secondary). Around 6000-7000 students were targeted annually for 10 years from 1999 to 2008. The project included planning, education sessions, blood collection, laboratory testing, and data processing, distribution of cards, data analysis and reporting.

The characteristic data for each student and test result were recorded. SPSS program was used for analyzing the data.

RESULT

The students screened during the ten years period were 60,000. The response rate to informed consent obtained from parents was 81-85%. Students free of beta thalassemia gene defect were 57,884 (96.5%). Table 1 and Figure 1 show the prevalence of beta thalassemia among these students.

Table 1: Prevalence of Beta Thalassemia among Students in Bahrain 1999-2008

Year	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
Disease	No and %	No and %	No and %	No and %	No and %	No and %	No and %	No and %	No and %	No and %
β thal Disease	5 0.09%	0 0.00%	3 0.05%	2 0.03%	3 0.06%	1 0.02%	1 0.02%	1 0.02%	0 0.00%	3 0.06%
β thal Trait	164 2.88%	212 3.72%	187 3%	219 3.72%	175 3.23%	212 3.40%	221 3.48%	231 3.64%	211 3.31%	265 5.05%
No of Student	**5685**	**5694**	**6244**	**5894**	**5418**	**6237**	**6358**	**6352**	**6376**	**5314**

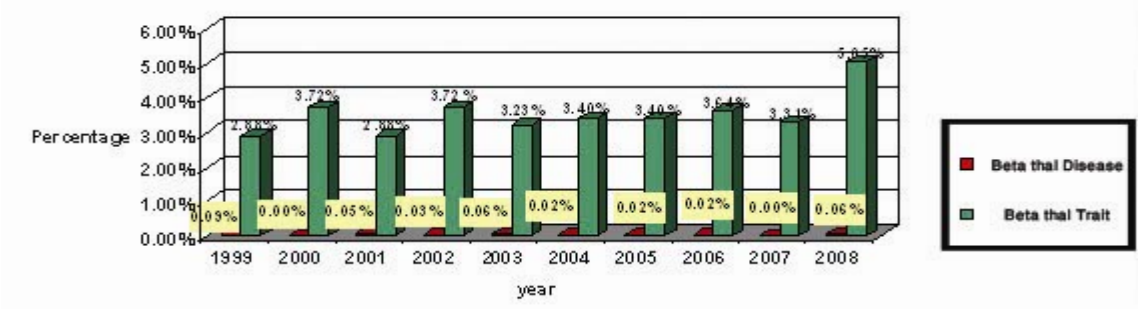

Figure 1: Beta Thalassemia Prevalence

Beta Thalassemia Major: The Number of beta thalassemia homozygous each year was 5, 0, 3, 2, 3, 1, 1, 1, 0, 2. Only five cases of beta thalassemia homozygous status, were detected in the first year.

The mean prevalence rate of beta thalassemia homozygous, SD, SE was 0.032%, 0 .029, and 0.00092 respectively. Significant difference was observed at 95% confidence (*p=0.004*). The lower and upper interval of difference was 0.0141 and 0.0559.

Prevalence of beta thalassemia Trait: The number of carriers for beta thalassemia during the years 1999-2008 was 164, 212, 187, 219, 175, 212, 221, 231, 211 and 265 students respectively.

The observed mean, SD, SE of beta thalassemia Trait were 3.5%, 0.614 and 0.194 respectively. The lower and upper interval of difference was 3.09 and 3.97.

Hb A2 in beta thalassemia trait ranged between 4% and 9%. Samples with lower figure of A2 (3.4-4) were suspected of having the gene and blood samples were directed for DNA studies to R/O heterozygosity.

Prevalence of beta thalassemia by Region in Bahrain: According to the result of 1999, some regions were observed to have a higher rate of this disease: Hidd 5.4%, Sitra 5.3%, Riffa 3.3% and Hamad town 3.35%. Other regions like Western region, Northern region, and Manama had only 2% prevalence rate. Similar results were observed during the subsequent years.

DISCUSSION

The prevalence of beta thalassemia in Bahrain was 3.5 % which was nearly the same figure obtained in the premarital study, the highest rate was found in Hidd 5.4% and Sitra 5.3%[24-25].

In the year 2008, there was a sudden increase in the frequency of beta thalassemia to 5%, which was not reported earlier. The cause of the rise of the carrier rate, and the future trend needs to be investigated.

The results of this study showed that there was a genetic heterogeneity in different regions in the kingdom of Bahrain. The Western area had the highest prevalence rate of sickle cell disease (25%) and it had the lowest prevalence rate of beta thalassemia (2%). In contrast with Al-Hidd region, it had the highest prevalence rate of beta thalassemia (5.4%) and the lowest prevalence rate of sickle cell disease (2.7%). Sitra region had the second highest rate for sickle cell disease (21%) and beta thalassemia (5.3%).

The malaria selection hypothesis could explain the higher rate of this disease in Sitra and Hidd, as they are small islands surrounded by water. The higher frequency rate of beta thalassemia in Riffa may be explained by the effect of migration from Hidd and Muharraq area (migration founder effect).

Internationally more than 500 mutations causing beta thalassemia have been characterized till date, the majority of which are non-deletional mutations[2,8,9]. In Bahrain, a previous study revealed 13 different beta thalassemia mutations, four different mutations accounted for 80% of all beta thalassemia alleles; sickle cell beta thalassemia was found in few cases[24-25].

CONCLUSION

The frequency of beta thalassemia disease in Bahrain is not high compared with many other Gulf countries such as UAE, Qatar and Kuwait.

Preventive measures such as health education, carrier screening and premarital counseling remain the best ways for lowering the incidence of these diseases, which might be reflected in financial saving, social benefits and health benefits.

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