ISSN 1996-3351

Asian Journal of Biological Sciences

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Asian Journal of Biological Sciences 6 (2): 124-130, 2013
ISSN 1996-3351 / DOI: 10.3923/ajbs.2013.124.130

A Demographic Approach for Understanding the Prevalence of β Thalassemia Patterns and Other Hemoglobinopathies: Selective Study in Chittagong City Perspective

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ABSTRACT

Beta thalassemia is an inherited autosomal recessive blood disease. In beta thalassemia and hemoglobinopathies, the genetic defect which could be either mutations or deletion results in reduced rate of synthesis or no synthesis of one of the globin chains that make up haemoglobin. Reduced synthesis or no synthesis of one of the globin chains can cause the formation of abnormal haemoglobin molecules, thus causing anemia, the characteristic presenting symptom of the thalassemia. The present study was undertaken with objective to study the occurrence and socio-demographic profile of hemoglobinopathies cases of Chittagong city. It may provide accurate data of population frequency that are necessary for the planning of control thalassemia in the highly populated eastern part of Bangladesh. This is a hospital record based prospected study that was carried out in two different General hospitals in Chittagong town where both hospitalized as well as out patients were included. The study was carried out from November 2011 to April 2012. A total of 485 suspected patients who were admitted in the hospitals were included in this study. Data was collected and then studied and analyzed properly. Analysis was done by employing Statistical Package for Social Science (SPSS Version 10.0) software package. Here the difference of male and female patients was found statistically insignificant ($p>0.05$). It was observed that, the onset of hemoglobinopathies was significantly high at neonatal to childhood period <1-10 years and the percentage was 51.70% ($p<0.05$). Here, the prevalence of HBE β Thalassemia patients (38.65%) was found the highest in percentage.

Key words: Hemoglobinopathies, thalassemia, genetic disorders, autosomal recessive, Globin Chains, Anemia, Bangladesh

INTRODUCTION

Thalassemia is one of the major autosomal recessive hereditary hemoglobinopathies prevalent in the world populations, particularly in Mediterranean belt, Far-eastern and South East Asian countries (El-Harth *et al*., 1999; Weatherall, 2001; Forget *et al*., 2000).

Mutations in the genes encoding α-chain and β-chain of haemoglobin are the primary cause of thalassemia resulting in the absence or inadequate synthesis of one of the globin chains (Weatherall, 1995). This anomaly in synthesis of globin chain can cause to the premature destruction of Red Blood Cells (RBCs), thus causing anemia and other secondary effects, the typical symptoms of thalassemia. In general, thalassemia can be classified into two pivotal forms, α thalassemia and β thalassemia by their clinical manifestations and genetic background. In α thalassemia, synthesis of the globin chain is impaired, whereas, in β thalassemia, synthesis of the β globin chain is affected. Beta thalassemia is the most frequent type of thalassemia which can be classified further into three cardinal forms; β thalassemia major, β thalassemia intermediate and β thalassemia minor/β thalassemia trait (Safizadeh *et al*., 2012). Individuals with β thalassemia major inherit two mutant β globin alleles (β^0); hence, synthesis of β chain is completely diminished with a consequence of the development of fatal anemia in early childhood if untreated. Intermediate β thalassaemic individuals carry mutation in one or both of the β globin genes whereas β thalassemic trait bears mutation in only one of the two β globin alleles (Talsania *et al*., 2011). Besides the commonest structural variant of haemoglobin E occurs globally due to point mutation in β globin gene which is innocuous in its heterozygous and homozygous states. Apart these when it interacts with β thalassemia alleles it produces an extremely common condition called Hb E/β-thalassemia. Hence it is becoming an increasingly important health burden in many parts of Asia (Rees *et al*., 1999). It has been estimated that, 5.2% of world populations are carrier of hemoglobinopathies and 2.7 per 1000 conceptions are affected. (Modell and Darlison, 2008). According to world health organization reports, approximately 3% of populations are carriers of beta thelassemia and 4% are carries of HbE in Bangladesh, which accounts 3.6 million of Beta thalassemia carriers and 4.8 million HbE/B-thalassemia carriers in the whole populations of Bangladesh (Khan, 1999). Moreover, a thalassemic child and its family undergoes through a socio-economic strain in whole community (Rahman and Rahman, 2003). Prenatal screening along with the status and distribution of thalassemia carriers provide better thalassemia management opportunity for the public health practitioners. In thalassemia prevalent developing countries carrier screening programs were successful in increasing the awareness about thalassemia among general mass (Xu *et al*., 2004). However, while Bangladesh is situated in a thalassemia prone region (Academisch, 2007), the country lacks in regional frequency data on the status and distribution of thalassemia carriers. Hence it is imperative to consider thalassemia as an important health issue in this country. So pragmatic steps are needed to be launched to reduce the birth rate of thalassemic children (Rahman and Rahman, 2003). Current study was conducted to observe the prevalence of different types of hemoglobinopathies, thalassemia and HbE among different age groups of Chittagong city. Chittagong-the port city is the second largest metropolitan city of Bangladesh where people from different regions of the country including a sizable number of tribal population lives which makes the city a suitable place to screen the distribution of thalassemia. Although the accurate statistics regarding the prevalence and degree of β thalassemia in this city is not well known, it seems to be escalating. If the disease continues to transfer vertically, it may become epidemic form in course of time. Since the effective treatment of this disease is unavailable, the best ways to prevent the disease are population screening, prenatal diagnosis and making mass awareness about this emerging epidemic. This study, therefore, provides a comprehensive data on the pattern and spectrum of hemoglobinopathies, β thalassemia and Hb E among the different age groups in Chittagong city.

MATERIALS AND METHODS

Study design: This is a hospital record based prospected study that was carried out in two different General hospitals in Chittagong town where both hospitalized as well as out patients were included:

- **Period of study:** November 2011 to April 2012
- **Study Subject:** A total of 485 suspected patients who were admitted in the hospitals were included in this study

Ethical consideration: Informed parental consents were taken before enrolling the patients into the study. The procedure was fully explained to the parents/local guardians and ensured that it would not hamper the treatment of the patients. Permission was also taken from the concerned departmental ethical committee in order to undertake the study.

Data analysis and presentation: Age, sex, type and pattern of Thalassemia were observed among the patients. Report of Haemoglobin electrophoresis of suspected patient's blood was collected from the hospital. Collected data was checked for its completeness, correctness. Editing and coding were done properly. Analysis was done by employing Statistical Package for Social Science (SPSS Version 10.0) software package. To compare mean values between groups, t-test was done as appropriate. $p = 0.05$ was considered as minimum level of significance. Data and results were presented in the form of tables and figures.

RESULTS

In this study a total 485 patients were enrolled (including the out patients) where 230 (47.42%) were male and 255 (52.58%) were female who were admitted to the two general hospitals in Chittagong town during November 2010 to April 2011. Among them 207 patients were directly linked with different types of Thalassemia and out of them 110 (53%) were male and 97 (47%) were female (Fig. 1). The difference of male and female Thalassemic patients was found statistically insignificant ($p>0.05$).

Out of total 207 patients, the individual percentage of HBE β Thalassemia, β Thalassemia Major, HBE Diseased, β Thalassemia Trait and HBE Trait in both male and female were calculated (Fig. 2). As was observed, HBE β Thalassemia was the most common one among all types of thalassemia amounting 38.65% of total 207 patients. this was followed by β Thalassemia Trait, which was 30.43%, was considerably more common than HBE trait (22.71%). The β Thalassemia major comprised 5.30%. It was little more than HBE Diseased (2.90%) which was least common. The differences between the thalassemia patients were found statistically insignificant ($p>0.05$).

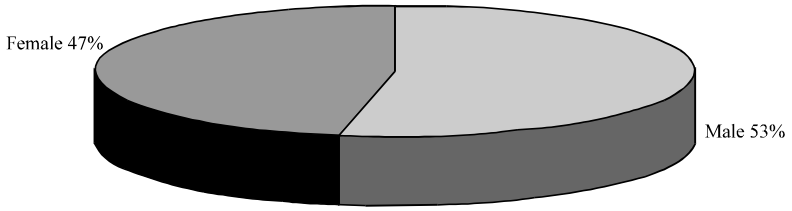

Fig. 1: Percentage of male and female thalassamia patients in this study. Gray shaded area indicated Male and Black shaded area indicated Female part, respectively

Table 1: Percentage of types of male and female thalassemia patients in different age groups

Types of diseases	Age groups (Years)	Male (%)	Female	Total
HBE β thalassemia	<1-10	33 (41.25)	23 (28.75)	56 (70)
	11-20	7 (8.75)	4 (5.00)	11 (13.75)
	21-30	5 (6.25)	3 (3.75)	8 (10)
	31-40	1 (1)	2 (2.50)	3 (3.5)
	>40	1 (1.25)	1 (1.25)	2 (2.5)
	Total	47 (58.50)	33 (41.50)	80 (100)
β Thalassemia major	<1-10	7 (63.63)	4 (36.37)	11 (100)
	11-20	0	0	0
	21-30	0	0	0
	31-40	0	0	0
	>40	0	0	0
	Total	7 (63.63)	4 (36.37)	11 (100)
HBE diseased	<1-10	2 (33.33)	1 (16.66)	3 (49.99)
	11-20	0	0	0
	21-30	0	2 (33.33)	2 (33.33)
	31-40	0	1 (16.66)	1 (16.66)
	>40	0	0	0
	Total	2 (33.33)	4 (66.65)	6 (100)
β Thalassemia trait	<1-10	11 (17.50)	8 (12.70)	19 (30.20)
	11-20	6 (9.52)	6 (9.52)	12 (19.04)
	21-30	2 (3.17)	12 (19.05)	14(22.22)
	31-40	3 (4.76)	7 (11.11)	10 (15.87)
	>40	6 (9.52)	2 (3.17)	8 (12.69)
	Total	28 (44.47)	35 (55.53)	63 (100)
HBE trait	<1-10	12 (25.53)	6 (12.77)	18 (38.30)
	11-20	7 (15)	4 (8.50)	11 (23.50)
	21-30	2 (4.25)	7 (15)	9 (19.25)
	31-40	4 (8.50)	2 (4.25)	6 (12.75)
	>40	1 (2.12)	2 (4.25)	3 (6.37)
	Total	26 (55.40)	21 (44.60)	47 (100)

Values in brackets are percentages

In the Table 1, the age, sex and type wise distribution of the thalassemia patients shows that, out of 80 HBE **β** Thalassemia patients 47 (58.50%) were male and 33 (41.50%) were female where in the age group between <1-10 years it was high 56 (70%). In case of **β** Thalassemia Major it was observed that, out of 11 patients 7 (63.63%) were male and 4 (36.37%) were female patients where it was only within the age group between <1-10 years. In case of HBE Diseased patients it was seen that, out of 6 patients 2 (33.33%) were male and 4 (66.65%) were female patients where maximum patients were in the age group between <1-10 years and it was 3 (49.99%). In case of **β** Thalassemia Trait individuals it was observed that, out of 63 cases 28 (44.47%) were male and 35 (55.53%) were female where within the age group between <1-10 years it was high 19 (30.20%). In HBE Trait cases it was seen that, out of 47 cases 26 (55.40%) were male and 21 (44.60%) were female where it was observed as above the incidence was high in the age group between <1-10 years and it was 18 (38.30%). This data suggested that among the all types of thalassemia the incidence of HBE **β** Thalassemia was high and in every case the incidence was high in the age group between <1-10 years.

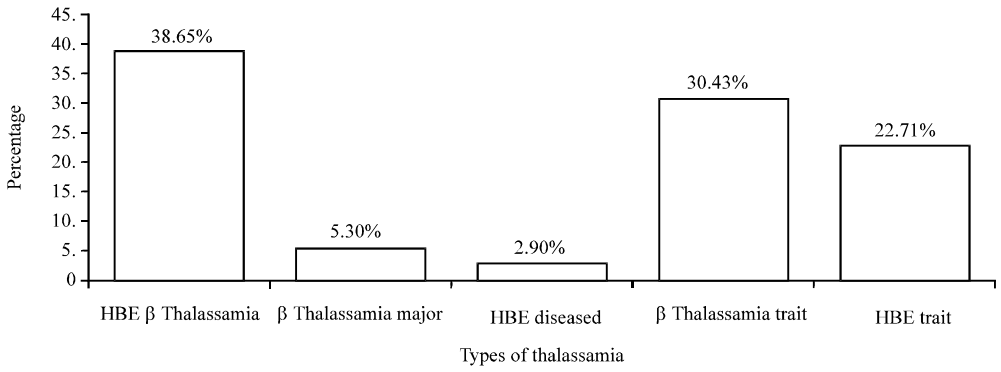

Fig. 2: Percentage of different thalassemia patients

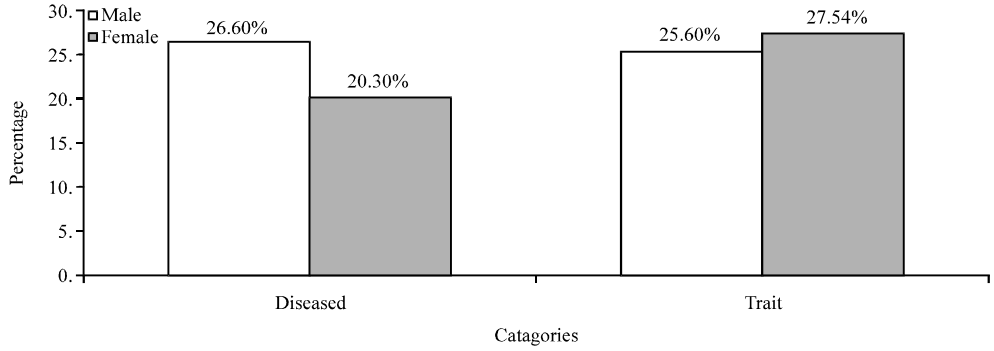

Fig. 3: Percentage of thalassemia diseased and trait patients in male and female

In this study out of the total patients (207) it was found that, there was 46.90% was Diseased where 26.60% were male and 20.30% were female. And 53.10% was Trait where 25.60% were male and 27.50% were female. This data stated that Thalassemia Trait were more than the Diseased patients (Fig. 3). The difference between Diseased and Trait of thalassemia patients were found statistically insignificant ($p>0.05$).

DISCUSSION

A total of 485 patients were enrolled in this study where 230 (47.42%) were male and 255 (52.58%) were female who were admitted to the two general hospitals in Chittagong town during November 2010 to April 2011. Among them 207 patients were directly linked with different types of Thalassemia and out of them 110 (53%) were male and 97 (47%) were female (Fig. 1). This data stated that in this study male patients were more in number than the female patients. Similar data was also found earlier by Wasi *et al.* (1985), Yagnik (1997) and Balgir (1996) and reported 65.5, 56 and 62.1% of male patients, respectively. Though the difference in the percentages was not significantly high but it might indicate that thalassemias are more common in male than in female in this region. This might be due to the gender bias among the parents of these ill children who seek medical care and are ready to spend more for their male children only. The difference of male and female patients was found statistically insignificant ($p>0.05$).

Among the enrolled patients, the individual percentage of HBE β Thalassemia, β Thalassemia Major, HBE Diseased, β Thalassemia Trait and HBE Trait in both male and female were (38.65),

(5.30), (2.90), (30.43) and (22.71%), respectively. Here, the percentage of HBE β Thalassemia patients (38.65%) was found the highest, the percentage of β Thalassemia Trait (30.44%) was found second highest and the percentage of HBE Trait (22.71%) was found comparably lower than HBE β Thalassemia and β Thalassemia Trait patients. But the percentage of β Thalassemia major and HBE Diseased patients found very few and they were 5.30 and 2.90% as a total, respectively. This data implies that HBE β Thalassemia is the most common form of thalassemias prevailing in this area. On the other hand, HBE diseased, is one of the hemoglobinopathies due to structural variants of haemoglobin, is the least common in this region. The differences between the thalassemia patients were found statistically insignificant ($p>0.05$). Previous studies at different part of Bangladesh also Showed similar results (Uddin *et al.*, 2012).

In the age, sex and type wise distribution of the thalassemia patients it was observed that, out of 80 HBE β Thalassemia patients 47 (58.50%) were male and 33 (41.50%) were female where in the age group between <1-10 years it was the highest 56 (70%). In case of β Thalassamia Major it was observed that, out of 11 patients 7 (63.63%) were male and 4 (36.37%) were female where it was only within the age group between <1-10 years. In case of HBE Diseased patients it was seen that, out of 6 patients 2 (33.33%) were male and 4 (66.65%) were female where maximum patients were in the age group between <1-10 years and it was 3 (49.99%). In case of β Thalassemia Trait individuals it was observed that, out of 63 cases 28 (44.47%) were male and 35 (55.53%) were female where within the age group between <1-10 years it was the highest 19 (30.20%). In HBE Trait cases it was seen that, out of 47 cases 26 (55.40%) were male and 21 (44.60%) were female where it was observed as above the incidence was highest in the age group between <1-10 years and it was 18 (38.30%). This data suggested that among the all types of thalassemia the incidence of HBE β Thalassemia patient, it was high and in every case the incidence was high in the age group between <1-10 years.

In this study out of the total patients (207) it was found that, there was 46.90% was Diseased where 26.60% were male and 20.30% were female. And 53.10% was Trait where 25.60% were male and 27.50% were female. This data suggested that Thalassemia Trait were more than the Diseased patients. The differences between Diseased and Trait of thalassemia patients were found statistically insignificant ($p>0.05$).

CONCLUSION

The considerably high prevalence of thalassemia in many Southeast Asian countries including Bangladesh is causing a major public health burden in this region. HBE β Thalassemia was the most common one among all types of haemoglobin formation disorders amounting 38.65% of total 207 patients. this was followed by β Thalassemia Trait (30.43%), HBE trait (22.71%), The β Thalassemia major (5.30%). However, proper population screening and making mass awareness can help to prevent thalassemia at least at a level so that it might not be epidemic in future. Prior conducting this research, the statistical data about prevalence of thalassemia in Chittagong city was inadequate. This study provides a comprehensive demographic data of pattern and spectrum of different hemoglobinopathies as well as β thalassemia among the different age groups in Chittagong city. It can be helpful to control thalassemia in this area and also provides a useful statistics for further research on thalassemia.

ACKNOWLEDGMENTS

Special thanks to Thalassemia Sheba Kendra, Chittagong, Chevron diagnostics and Sensiv for their cooperation during the study.

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