Clinical and Socio-demographic Pattern of Beta Thalassaemia in Bangladesh

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ABSTRACT

Background: Beta thalassaemia is one of the most prevalent haemolytic disorders worldwide which poses serious economic burden to the society. **Objective:** Aim of the Study is to see clinical and demographic pattern of Beta thalassaemia which will help the concern authorities to figure out the problem. **Method:** This cross-sectional descriptive study was carried out to determine the socio-demographic and clinical characteristics as well as to find out the existence of other co-morbidities among the β -thalassaemic patients includes 101 subjects. The study was carried out at the Department of Haematology, Bangabandhu Sheikh Mujib Medical University (BSMMU) from June 2010 to January 2011. Data were collected from 3 tertiary care hospitals of Dhaka city. **Results:** Almost all the patients were young, age ranged from 10 to 32 years with the mean age of 16. Male to female ratio was almost equal (52.5:47.5) and 80% of the respondents were Muslims. Forty-five (45%) percent of patients were illiterate and 37% respondents had primary education only. Nearly 90% were unmarried and majority had a family of 6-8 members. Forty-seven percent (47%) of patients had monthly income 5000-7000 BDT and 56% of the respondents were unemployed. The major clinical features were pallor (72.3%), palpitation (60%), and breathlessness (52.5%). The haemoglobin (Hb) concentration ranged from 7gm/dl to 12gm/dl and 57% patients had an Hb concentration of 8 gm/dl. Sixty percent of the respondents had jaundice with majority had enlarged spleen (86%) and some had enlarged liver too (23%). Ninety percent (90%) respondents had co-morbidities like- arrhythmias, recurrent infections and skin pigmentations among which arrhythmia is more prevalent (54%). **Conclusion:** The clinical symptoms along with other co-morbidities relate the finding of unemployment which is also a burden for their family as well as the society.

Keywords: comorbidity, educational level, monthly income, thalassaemia.

Introduction

Thalassaemia is the most common genetic disorder worldwide that causes a reduction in the rate of synthesis of one or more types of normal haemoglobin (Hb) polypeptide chain.¹ It is classified in two main genetic

groups: α -thalassaemia that affect synthesis of α chain, and β thalassaemia that affect synthesis of β chains.^{2,3} The genetic mutation may also result in the synthesis of a structurally abnormal haemoglobin along with reduced

haemoglobin production.² Clinically, they are classified as – (1) thalassaemia trait or carrier, usually asymptomatic, (2) thalassaemia major, regular transfusion dependant, and (3) in between them is thalassaemia intermedia. As the disease follows the Mendelian rules of inheritance, the children of a carrier couple have a risk to have thalassaemia major.⁴

The symptoms of beta thalassaemia major start to develop between the age of three and six months that include features of anaemia, medullary expansion and extramedullary haemopoiesis. Moreover, they may also have the features of complication of iron overload as they need repeated blood transfusion and also there is increased iron absorption from gut.⁴⁻⁷ Cardiac complications of iron overload represent the main determinants of survival in these patients.⁸⁻¹⁰ Iron chelation therapy can eliminate or reduce cardiac and other complications but compliance with it is very deficient.¹¹ Musculo-skeletal problems occur due to anatomic proximity of bones and joints to the active centres of haematopoiesis.¹² These include fractures, premature epiphyseal fusions and thalassaemic osteo-arthritis, especially in the more severe variety of beta thalassaemia major.¹³⁻¹⁷

Thalassaemia is prevalent more often in the Middle East, India, South East Asia (including southern China, Thailand and Malaysia) and parts of Africa and the southern Mediterranean with a reported carrier rates ranging from 2% to 30%.¹⁸⁻¹⁹ The WHO estimates that about 7% of world populations are carriers.¹⁹ About 300000-500000 children are born each year with the severe homozygous states of this disease.²⁰ The estimated prevalence of beta thalassaemia is 16% in Cyprus, 3-14% in Thailand and 3-8% in India, Pakistan, Bangladesh and China.²¹ Prevalence is low in African black people (0.9%) and Northern Europe (0.1%).²²

In Bangladesh, there is no consensus report regarding the carrier status of hereditary Hb disorders. A conservative World Health Organization report estimates that 3% of the populations are carrier of β -thalassaemia in Bangladesh with 0.106 affected births per thousand; it means >2,000 thalassaemia children are born every year in Bangladesh.²³ However, precise data related to socio-demographic characteristics and clinical status of β -thalassaemic patients are scarce. Therefore, this study was carried out to identify the socio-demographic characteristics and clinical status of β -thalassaemic patients and any co-morbidity of these patients accordingly. The results of this study may help to develop a strategy of helping the policy makers to take corrective measures for helping the thalassaemic patients.

Materials and Methods

This was a descriptive cross-sectional study conducted at the Department of Haematology, BSMMU, Dhaka, Bangladesh, from June 2010 to January 2011. Ethical permission was taken from the institutional review board of BSMMU. According to the statistical calculation, 101 β thalassaemic patients of both sexes from outpatient and in-patient Haematology Departments of Dhaka Medical College Hospital, Mitford Hospital and BSMMU hospital of Dhaka city were enrolled in the study. The patients were selected purposively irrespective of age and sex. After taking informed written consent, data were collected with a structured questionnaire. The relevant investigations such as Hb% and, Hb-electrophoresis were reviewed, and clinical examinations done. Their socio-demographic data such as age, sex, religion, educational status, occupation, marital status, monthly family income and number of family members were taken. Their clinical features such as pallor, palpitation, breathlessness, haemoglobin, jaundice, size of the spleen and liver were evaluated. Finally, other co-morbidities such as arrhythmia, recurrent infection and pigmentation were also searched for. All the data were analysed with the help of SPSS windows program version 18.

Results

Among 101 patients, males (52.5%) were slightly outnumbered than the females (47.5%) with an age range from 10 to 32 years. The mean (mean $\pm$ SD) age of the patients was 16.60 ± 5.09 . Four-fifth of the respondents (79%) were below the age of 20 with major distribution in 10-14 years age group (44.6%). Prevalence of β -thalassaemia decreases with increasing age of the patients. Most of the patients were Muslims (80.2%) followed by Hindus (17.8%), Buddhists (1%) and Christians (1%). Regarding the education level of the patients, only one patient studied at higher secondary level, 44.6% patients were illiterate being the largest group among the patients, 37.6% had completed their primary education and 16.8% completed their secondary education. Majority of the respondents were unemployed (56.4%) followed by students (35.6%), housewife (6.9%) and only one businessman. Majority of the respondents were unmarried (93.1%). The range of income varies from 3,000 BDT to 34,000 BDT with mean income of 8519.80 BDT, but 80% respondents had the family income less than 9000 BDT per month. The largest (46.5%) income group was within 5001 to 7000 BDT. Among them, 55.4% patients had a family of 6 to 8 members, 31.7% had 3 to 5 members, 11.9% had 9 to 11 members and one patient had a family of 12 members. (Table I).

Table I: Demographic characteristics of study subjects. (n:101)

Demographic characteristics	
Age in years (mean $\pm$ SD)	17 $\pm$ 5
Age groups; n (%)	
10-14 Years	45 (44.6)
15-19 Years	35 (34.7)
20-24 Years	10 (9.9)
$\geq$ 25 Years	11 (10.9)
Gender; n (%)	
Female	48 (47.5)
Male	53 (52.5)
Religion; n (%)	
Muslim	81 (80.2)
Hindu	18 (17.8)
Buddhist	1 (1.0)
Christian	1 (1.0)
Educational status; n (%)	
Illiterate	45 (46.6)
Primary	38 (37.6)
Secondary	17 (16.6)
Higher secondary	1 (1.0)
Occupation; n (%)	
Student	37 (36.6)
Unemployed	58 (57.4)
Housewife	5 (5.0)
Businessman	1 (1.0)
Marital Status; n (%)	
Married	7 (6.9)
Unmarried	94 (93.1)
Family member; n (%)	
3-5	32 (31.7)
6-8	56 (55.4)
9-11	12 (11.9)
>11	1 (1.0)
Monthly income in BDT; n(%)	
$\leq$ 5000	15 (14.9)
5001-7000	47 (46.5)
7001-9000	19 (18.8)
>9000	20 (19.8)

All the patients had pallor, 72.3% had mild and 27.7% had moderate pallor. Enlarged spleen was present in 86% of the patients. Jaundice and palpitation were present in 60% and 60.4% of the patients, respectively. Hepatomegaly was present in 24% of patients. Most of the patients (90%) had other diseases or complication e.g., recurrent

infections in 28.7% and hyperpigmentation in 12.9% of the study patients. Majority (85.1%) of the patients had the haemoglobin level between 9-7 gm/dl. Only 14.9% respondents had Hb level >9 gm/dl. (Table II)

Table II: Clinical and laboratory findings of study subjects. (n:101)

Major Clinical findings	n (%)
Pallor	
- Mild	73 (72.3)
- Moderate	28 (27.7)
Palpitation	61 (60.4)
Breathlessness	53 (52.5)
Jaundice	61 (60.4)
Splenomegaly	87 (86.1)
Hepatomegaly	24 (23.8)
Associated Co-morbidity	n (%)
Recurrent infection	29 (28.7)
Pigmentation	13 (12.9)
Haemoglobin level (gm/dl)	n(%)
7	12 (11.9)
8	57 (56.4)
9	17 (16.8)
10	5 (5.0)
11	8 (7.9)
12	2 (2.0)
Mean $\pm$ SD	8.47 $\pm$ 1.15

Discussion

This study revealed that most of the cases of beta thalassaemia in Bangladesh are found in teen aged, less educated, Muslim, unemployed males with low socio-economic status. All the patients were anaemic, and splenomegaly was found in majority of the patients. Besides these, recurrent infection and skin pigmentation also developed as the consequence of thalassaemia.

Males are slightly outnumbered than the female which is reflected male female ratio published by Bangladesh Bureau of Statistics (BBS) (106:100) and in Bangladesh this disease is prevalent in same intensity among the both sexes.²⁴ In Northern India, males were affected more than females (71.4% vs 28.1%; male vs female).¹⁶

Teen aged peoples were more affected justifying that beta thalassaemia is a disease of childhood. This result was similar to a study done in Northern India where the mean age of the patients was 17.2 years.¹⁶ Similar result

was also revealed in a study by Khan in 1999.²³ Improved treatment and investigation facilities may be the cause behind it. The religion of the patients corresponds the national data of BBS (88.3% Muslims, 10.5% Hindus and 1.2% Buddhist and Christian).²⁴

The lower level of educational status in this study might be due to the costly and hazardous treatment procedure that keeps the thalassaemic patients away from taking institutional education. However, a study in India showed that the patient's family had a higher literacy status to graduate level.¹⁶ Only 7% were married and, notably, all were female.

About half of the respondents (56%) were unemployed which may be related to lack of body fitness associated with disease. The range of income of the respondents was from 3000 BDT to 34000 BDT with mean income of 8519 BDT which was much lower in relation to treatment cost of thalassaemia patients. About 87% respondents had a family member of 3-8 and the rest had more than 8. So, number of family members was also more in respect to monthly family income.

Anaemia was the ubiquitous clinical feature of all the patients. Other symptoms such as palpitation and breathlessness were also found in more than 50% of the patients which were due to the less haemoglobin concentration as well as decrease O₂ carrying capacity of blood. Around 66% had a haemoglobin concentration less than 8 gm/dl which was much lower than actual recommended need. Several other studies also revealed similar concentration of haemoglobin.²³⁻²⁴

About 60% patients had jaundice, 86% had splenomegaly and 23% had hepatomegaly. Actually, among all the respondents, 23% developed both hepato-splenomegaly. These findings coincide with classical features of ineffective erythropoiesis.² About 90% patient developed co-morbidity with arrhythmia, recurrent attack of infection and skin hyperpigmentation.

Conclusion

Though this study was carried out in small number of patients, the findings were remarkably interesting revealing poor socio-economic and low educational status in the thalassaemic patients. The clinical symptoms along with co-morbidities relate the finding of unemployment which is also a burden for their family as well as the society. All the findings in the study underscore the importance of thalassaemic patient's care and pre-marital screening and counselling. Considering the findings, this study recommends to the policy makers, preventive medicine specialists, future researchers and thalassaemic

individuals to improve the family income and literacy rate. The study also recommends establishing special care and diagnostic facilities, rehabilitation centres for the patients, and providing genetic and marriage counselling to avoid the unexpected birth of thalassaemic patients. Furthermore, research with large sample size should be carried out in a broader aspect and take positive effort for the prevention of β -thalassaemia.

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