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Demographic and History Related Treatment Status of Thalassaemia Patients in a Tertiary Hospital in Bangladesh

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ABSTRACT

Background: *Thalassaemia is now effectively treated with adequate blood transfusion and iron chelation. The disease processes itself and iron overload from blood transfusions together produce multiple complications. Due to increased life expectancy of thalassaemia patients it is important to point out their demographic profiles that may vary due to numerous causes.* ***Objective:*** *Aim of the study is to find out demographic profiles of thalassaemia patients. Secondary objectives are to categorize the demographic characteristics according to three clinical thalassaemia types, and to assess history related treatment parameters like transfusion status, splenectomy, iron chelation and compliance to proper treatment.* ***Methodology:*** *Three clinical types of thalassaemia are assessed in Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh between July 2016 to July 2017. Total 109 subjects with thalassaemia are included in the study according to the inclusion criteria. They are categorised as thalassaemia major (33 subjects), thalassaemia intermedia (34 subjects) and thalassaemia minor (42 subjects). The patients are asked questions regarding demographic status and some clinical histories through a simple data collection sheet after proper consent. After compilation of the data, statistical analysis was done accordingly.* ***Results:*** *Mean (±SD) age of thalassaemia major patients is 22.73±11.62, thalassaemia intermedia 22.09±12.55 and thalassaemia trait 20.50±11.37 with nearly equal gender distributions. 27.27% subjects among thalassaemia major, 32.35% subjects among thalassaemia intermedia and 30.95% subjects among thalassaemia minor are students. The percentage of participants having formal education up to primary level in Bangladeshi education system is 51.52% in thalassaemia major, 47.06 % in thalassaemia intermedia and 57.14 % in thalassaemia minor. Most of the subjects are Muslims and over 75% of the subjects came from outside the Dhaka city. On an average all categories of thalassaemia subjects received 42.96±73.22 (mean ± SD) blood transfusions in their lives so far. The average of life time transfusions are 105.33±87.36 (mean ± SD) in the patients of thalassaemia major, 33.85±61.62 (mean ± SD) in the intermedia and 0.76±2.37 (mean ± SD) in the minor. Iron chelation are received by 48.48% of thalassaemia major and 17.64% of thalassaemia intermedia patients at some point of their treatment. Splenectomy is done in 5 (15.15%) of thalassaemia major and 2 (5.8%) of thalassaemia intermedia patients. Before enrolment in the study 9.09% thalassaemia major, 50% intermedia and 83% minor subjects never visited haematologists or haematology outpatient services.* ***Conclusion:*** *Transfusion frequency is high with inadequate iron chelation and young people with thalassaemia are in need of adequate specialized treatment in their local area in Bangladesh.*

Key Wards: Thalassaemia, Demography, Treatment status.

Introduction

Human beings suffer from the legacy of disease of their ancestors as many inherited disorders run within the family tree. Thalassaemia is one of them with immense importance. This disease has prevailed for over thousands of years in the history of mankind. It is also known as Mediterranean anaemia. Skeletons with severe 'porotic hyperostosis' are found in several tombs across Sicily and Sardinia which are assumed to be due to severe congenital haemolytic condition i.e. thalassemia.[1] In the coming 20 years, there will be an estimated 900,000 births of clinically significant thalassemia disorders.[2]

Thalassemia inherits according to Mendel's laws.[3] There is deficient production of one or more of the globin subunits of haemoglobin resulting in reduced or absent synthesis of normal haemoglobin. According to World Health Organization (WHO) at least 3% of Bangladesh populations are carriers of beta thalassaemia. Approximately 6000 thalassaemic children are born each year in Bangladesh with thalassaemia.[4] A single centre study in Bangladesh has observed the frequency of hereditary haemoglobin disorders as Hb-Eβ Thalassemia by 68.50%, β Thalassemia major by 31% and Hb E disease by 0.5% and no α-thalassemia.[5]

Thalassaemia can be classified in various ways depending on genetic or clinical status. According to the type of globin chain that is produced in reduced amount, thalassaemia can be classified into two major categories- the alpha and beta thalassaemia. The clinical severity thalassaemia is of three types.[6] It includes thalassemia major, thalassemia intermedia and thalassemia minor.[7] This classification usually indicates the necessity of blood transfusion along with other clinical parameters.

Thalassaemia has a broad spectrum of presentations starting from asymptomatic to life threatening conditions. The β thalassemia major is treated with life-long mandatory regular transfusion therapy to maintain the normal life but, as a double-edged sword, it also results in toxic iron overload.[8,9] Therefore, chronic hypoxia and uncontrolled iron overload possess serious clinical consequences of morbidity and mortality. Almost every system of the body is damaged in these conditions, most commonly the liver, heart and endocrine glands.[8]

Advances in the treatments of thalassaemia have improved prognosis. The wide spread availability of safe transfusions and iron chelation have dramatically prolonged the life of these patients.[10] Now a days, majority of patients of thalassaemia survive to adulthood.[11]

In a middle-income country like Bangladesh, it is important to study the demographic and investigation dependent factors related to thalassaemia. It may pave a way to find out obstacles for prevention and fruitful treatment of thalassaemia and go for a proper solution and resource allocations.

Materials and methods

This was a cross-sectional observational study done in the Department of Haematology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbagh, Dhaka, Bangladesh in a period between July 2016 to July 2017 after having necessary ethical approval. The objectives are explained to each participant or the guardian. Considering power of study 0.80, standard deviation of means 2.16, standard deviation 6.0, effect size0.36, alpha 0.05, group number three for three types of thalassaemia; the minimum total number of samples for this study is being calculated to be 78 by automated software. So, the average group sample size is 26. Convenient sampling method used. After assessing haemoglobin electrophoresis or DNA analysis, data collection sheet (attached in appendix) is filled up by subjects or their guardian after proper consent. Any thalassaemia subjects at or above one-year age are selected for the study. They are categorized in three clinical types -thalassaemia major who had received eight or more transfusions during last twelve months, thalassaemia intermedia who had received less than eight transfusions during last twelve months and thalassaemia minor who had received no transfusions during last twelve months.[12] After compilation of data, statistical analysis is done accordingly.

Results

Demographic profiles: Total 109 subjects are included. Among them, 33(30.27%) are thalassaemia major, 34 (3.9%) are thalassaemia intermedia and 42(38.53%) are thalassaemia minor. (Table I)

Gender distribution: Among the thalassaemia major, 19 (57.6%) patients are male and 14 (42.4 %) are female. Among 34 patients of thalassaemia intermedia, 19 (55.9 %) are male and 15 (44.1%) are female. Among the 42 patients of thalassaemia minor, 18 (42.9%) are male and 24 (57.1%) are female. (Table I)

Age distribution: Mean (±SD) age and standard deviation of subjects of thalassaemia major patients was 22.73±11.62, thalassaemia intermedia 22.09±12.55 and thalassaemia trait 20.50±11.37. (Table I)

Religion: All subjects in thalassaemia major and thalassaemia intermedia are Muslims. Only 11.9 % subjects in thalassaemia minor are not Muslims. (Table I)

Educational status: Educational status of thalassaemia major group reveals 6.06% illiterate, 9.09% can only sign their name, 51.52% completed primary school level of education, 18.18% achieved Secondary School Certificate, 6.06% achieved Higher Secondary School Certificate, 36.06% had Diploma and 03% are of pre-school age. In thalassaemia intermedia group educational status reveals; 5.58% can sign only, 47.06% primary school level, 23.53 % Secondary School Certificate, 8.82 % Higher secondary School Certificate, 2.94 % post graduate and 11.76% pre-school age. Educational level of thalassaemia minor subjects shows; 2.38 % illiterate, 57.14 % primary, 7.14% Secondary School Certificate, 11.90 % Higher secondary School Certificate, graduate 14.29 % and pre-school age 11.90.2%. (Table I)

Table I: Demographic characteristics of subjects among clinical types of thalassaemia.

Characteristics	Type of Thalassaemia major	intermedia	minor
Age in Years; Mean±SD	22.7±11.6	22.09±12.55	20.5±11.38
Gender (Male: Female)	19:14	19:15	18:24
Religion; Muslim: Others	33: 0	34:0	37:5
Education; n (%)			
Illiterate	2 (6.06)	0 (0.0)	1 (2.38)
Can sign only	3 (9.09)	2 (5.88)	0 (0.0)
Primary	17 (51.52)	16 (47.06)	24 (57.14)
SSC	6 (18.18)	8 (23.53)	3 (7.14)
HSC	2 (6.06)	3 (8.82)	5 (11.90)
Graduate	0 (0.0)	0 (0.0)	6 (14.29)
Post graduate	0 (0.0)	1 (2.94)	0 (0.0)
Preschool	1 (3.03)	4 (11.76)	5 (11.90)
Diploma	2 (6.06)	0 (0.0)	0 (0.0)
Occupation; n (%)			
Service holder	4 (12.12)	0 (0.0)	5 (11.90)
Business	2 (6.06)	4 (11.76)	3 (7.14)
Housewife	6 (18.18)	9 (26.47)	11 (26.19)
Student	9 (27.27)	11 (32.35)	13 (30.95)
Farmer	1 (3.03)	1 (2.94)	0 (0.0)
Others	10 (30.30)	8 (23.53)	9 (22.0)

Professional category: Professional category of thalassaemia major group reveals; 12.12 % service holder, 6.06% business man, 18.18% housewife, 27.27% student, 3.03% farmer and 30.3% other professions. In thalassaemia intermedia group; 11.76% are businessman, 26.47% housewife, 32.35% student, 2.94 % farmer and 23.53% from other professions. In thalassaemia minor group; there are 11.9% service holder, 7.14% businessman, 26.19% housewife, 30.95% student and 22.0 % other professions. (Table I)

Address: Most of the patients of all clinical types came from outside Dhaka city i.e. 84.4% of thalassaemia major, 88.2% of thalassaemia intermedia and 75% of thalassaemia minor patients. Figure four showing percentage of subjects coming from Dhaka district and from outside Dhaka district in three clinical types of thalassaemia. (Figure 1)

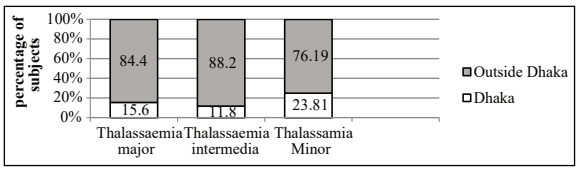

Figure 1: Percentage of subjects coming from Dhaka district and outside Dhaka district in clinical types of thalassaemia.

On an average all thalassaemia subjects received 42.96±73.22 (mean ± SD) times blood transfusions in their lives so far. However, among the thalassaemia major subjects the mean (± SD) of total number of transfusions in life is 105.33±87.36. It is 33.85±61.62 (mean ± SD) in thalassaemia intermedia and 0.76±2.37 (mean ± SD) in thalassaemia minor. total 15.5% (5) subjects with thalassaemia major have been splenectomised while in thalassaemia intermedia the percentage is only 5.8% (2) and none in thalassaemia minor. Around 48% thalassaemia major patients had access to iron chelation in some point of their treatment but it is about 17.64% in thalassaemia intermedia. Only 9.09% thalassaemia major subjects strictly followed haematologists' prescriptions or haematology outpatient department (OPD) advices. It is 8.82% in thalassaemia intermedia and 4.76% in thalassaemia minor. Alarmingly 9.09% thalassaemia major, 50% thalassaemia intermedia and 83% thalassaemia minor subjects never consulted a haematologist or haematology OPD. Rest of the percentage are on and off regarding proper consultation from haematologists.

Discussion

In this study among total 109 subjects 33 subjects are in thalassaemia major, 34 in thalassaemia intermedia and 42 in thalassaemia minor group. A study in Sri Lanka has found 68.7% thalassaemia major, 5% thalassaemia intermedia, 20.3% E Beta thalassaemia and 0.9% beta thalassaemia trait.[13] A North American study has found 54% thalassaemia major, 15% thalassaemia intermedia and 13% E-Beta thalassemia among the subjects. In India 91.1% thalassaemia major and 7.1% E-Beta thalassaemia are found in a

study.[14] Definition of thalassaemia types in various studies, sample size, different geographic locations, birth rate, education, migration, awareness in thalassaemia control programme can give rise to the differences of percentages of thalassaemia types.

The mean age of the subjects of thalassaemia major, intermedia and minor are 22.73±11.62, 22.09 ±12.55 and 20.50±11.37 years respectively. In a study at different cities in Iran has found that mean age of thalassaemic subjects ranging from 11.39 years to 30.72 years varying according to type of thalassaemia, sex, location etc.[15] An Indian study showed mean age of 127 months ranging from 18 to 492 months. The variation is dependent on the inclusion criteria.[16]

Among the thalassaemia major group 57.6% patients are male and 42.4 % are female. In thalassaemia intermedia group 55.9 % male and 44.1% female while in the thalassaemia minor category 42.9% male and 57.1% female subjects are found. In a study; 52.8% female, 47.2% male in adult population and 46.9% female, 53.1% male in paediatric population among thalassaemia major group was found in Italy.[17]

Among the total study subjects, according to occupation majority represented are students. The percentage is 27.27% in thalassaemia major, 32.35% in thalassaemia intermedia and 30.95% in thalassaemia minor. Housewives are the second highest as profession showing 18.18%, 26.47% and 26.19% respectively in thalassaemia major, thalassaemia intermedia and thalassaemia minor. Unfortunately, most of the students and housewives in Bangladesh are economically dependent resulting in poor access to proper treatment of thalassemia. A study in North America has shown that 70% of adults are employed of which 67 % reported working full time.[14] A previous study in Baghdad showed that among the thalassaemic subjects there are 16% student, 7% employed,1% retired, 24% freelancer and 36% house wives.[18]

In this study the majority of subjects are educated upto primary educational level as 51.52% in thalassaemia major, 47.06 % in thalassaemia intermedia and 57.14% in thalassaemia minor patients belonging to primary level of education. In a study in North America, 60% college degree, 14% post college degree and 82% school going children at their expected class are found.[14] In another study in Iran, 60% thalassaemia major, 50% thalassaemia intermedia and 70% sickle beta thalassaemia subjects are at less than high school education and in Iran shahr, the poorest city in Iran, had 54.5% illiteracy rate.[19] In a study in India, it was 54.7% in primary school level, 29.2% high school level and 14.6% graduate. Economic standards and strength of social welfare services may contribute to the differences among studies.[16]

Most of the subjects are Muslims in all groups as Bangladesh is a Muslim majority country. So as an Indian study showed 74.4% Hindu, 20.5% Muslim and 5% other religious groups in thalassaemia subjects.[20]

In this study 75% or more of the subjects in all groups are from outside Dhaka city. It is a major problem that people had to travel a long way for treatment of thalassaemia giving rise to poor compliance to treatment.

This study revealed that average transfusion received by the subjects through their life is 42.96±73.22. The thalassaemia major subjects received average 105.33±87.36 (mean ± SD) transfusions in their life. In intermedia and minor group, it was 33.85 ±61.62 (mean ± SD) and 0.76 ±2.37 (mean ± SD) respectively. Thalassaemia patients received an average total number of 149 ± 103.4 units of red blood cells in a study in USA.[21]

In this study, 48.48% thalassaemia major and only 17.64% thalassaemia intermedia subjects received iron chelation at some point of their treatment which was 100% in studies in India and Iraq.[22,18] This is a major problem in Bangladesh as we are not able to provide proper iron chelation and as a result complications are uncontrolled.

In this study 15.15 % of thalassaemia major subjects had splenectomy and the number is only 2(5.8%) in thalassaemia intermedia and obviously zero in thalassaemia minor. It was 34% in a study done in Iraq, 31.7% in Iran, 47.4% in India and 23% in another study carried out in Iraq.[18,19,22,23]

Unfortunately, only 9.09% subjects of thalassaemia major, 8.82% of thalassaemia intermedia and 4.76% of thalassaemia minor have strictly followed haematologist or haematology OPD prescriptions. The irony is; 9.09% thalassaemia major, 50 % intermedia and 83% minor subjects have never visited haematologists or haematology OPD before being enrolled in the study. Rest of the subjects are irregular regarding consulting and following haematologists' prescription. Regarding total compliance specific data are not available. But a study in Iran has shown 85.1% good compliance regarding iron chelation which is definitely better than the rate in present study.[19] This poor compliance of treatment indicates that subjects with thalassaemia are not able to maintain standard quality of life in Bangladesh.

Conclusions

Assessment of some demographic and history related clinical characteristics in three clinical types of thalassaemia at a cross section of time is momentous observation of this study. This is like a single step as a single centre study but a giant leap for inviting future studies regarding thalassaemia in Bangladesh.

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