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Original Article

Pattern of Thalassemia and other Hemoglobinopathies in Sylhet, Bangladesh

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Abstract:

Background: Thalassemias and hemoglobinopathies are the most common inheritant hemolytic congenital disorders in Bangladesh.There is no well precise validated data available about the prevalence of Thalassaemia and related hemoglobin disorders in Bangladesh.

Method: This retrospective, cross-sectional observational study was carried out in Dr.Benzamin's Pediatric Liver Research Centre and Nutrition Clinic, Sylhet, Bangladesh. We reviewed the data record software of the hematology section of the Popular Diagnostic Centre and Mount Adora Hospital, Sylhet, from March, 2022 to July, 2023 and collected all the Hemoglobin electrophoresis reports.A total of 783patients data were evaluated, and the data were entered into Microsoft Excel and analyzed by Statistical Package for Social Sciences (SPSS) software version 22. The aim of the study was to identify the pattern of Thalassemia and other Hemoglobinopathies in Sylhet, Bangladesh.

Result: Out of 783 patients, 291(37.2%) were male, 492 (62.8%) female, pediatric population that is under 18 years 387 (44.4%) and 18 years or more 396(50.6%). Among these, 262 (33.46%) reports showed Hemoglobinopathies. Before 6 months, 10 patients were advised for Hb electrophoresis. Overall, in study population, most common hemoglobinopathies were Beta thalassemia trait (118, 15.1%), followed by Hb E trait (45, 11.6%), Hb E Beta thalassemia (34, 4.43%), Beta thalassemia major (8, 0.9%). Same trend followed in pediatric age group, Beta thalassemia trait (53, 13.7%), Hb E tarit (84, 10.7%), Hb E Beta thalassemia (28,7.24%), Beta thalassemia major (8, 1.8 %). In adult age group, Beta thalassemia trait (65, 16.4%), Hb E tarit (39, 9.8%), Hb E Beta thalassemia (6, 1.5%%).

Conclusion: Hemoglobinopathies among the anaemic population of Sylhet is very common. Beta thalassaemia trait, Hb E tarit, Hb E Beta thalassemia, Beta thalassemia major are common variants. So, during anaemia evaluation, hemoglobinopathies should be kept in mind.

Key words: Hemoglobinopathies, Thalassemia, Hb electrophoresis.

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Introduction:

Thalassemias and hemoglobinopathies are the most common inheritant hemolytic congenital disorders in Bangladesh.[1]Haemoglobinopathies denote genetic defects collectively affecting either haem structure or synthesis.[1]They are divided into two categories.

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First are those that affect the structure of hemoglobin (Hb) and produce abnormal Hb structural variants usually due to substitution of one amino acid by another for example hemoglobin C, S, D and E. Second type, called thalassemia, primarily affects the rate of synthesis of one of the globin chains causing an imbalance in the production of Hb subunits.[2]

Now a days, Hemoglobinopathies have emerged as a global public health concern. An estimate of World Health Organization shows that 7% of the world population are a carrier for hemoglobin disorders and 300,000 to 500,000 children are born with clinically significant hemoglobin disorders every year.[1] Alarming scenario is, 80% of these born in low income countries.[3] There is

no well precise validated data available about the prevalence of Thalassemia and related hemoglobin disorders in Bangladesh. It's presumed from multiple studies that around 3% and 4% of Bangladeshi population are carrier of β Thalassemia and Hb E respectively.[4] Hemoglobinopathies are important cause of morbidity and mortality worldwide. Birth of a single child with a hemoglobin disorder place a large burden to the patients, their families, and even their communities and causes enormous psychological trauma along with financial strain for treatment especially in developing countries like Bangladesh. The patients suffering from beta-thalassemia major and HbE/beta-thalassemia do not survive for more than 5 years without blood transfusion.[5] Worldwide 3.4% of mortality rate in children under 5 years age are due to Hb disorders.[1]

Methods:

This retrospective, cross-sectional observational study was carried out in Dr. Benzamin's Pediatric Liver Research Centre and Nutrition Clinic, Sylhet, Bangladesh. The aim of the study was to identify the pattern of Thalassemia and other Hemoglobinopathies among the anaemic populations of Sylhet, Bangladesh. We reviewed the data record software of the hematology section of the Popular Diagnostic Centre and Mount Adora Hospital, Sylhet, from March, 2022 to July, 2023 and collected all the Hemoglobin electrophoresis reports of patients who presented to physician/ hospital with anemia. Hemoglobin electrophoresis was done by following method: 2 ml of venous blood was collected in a tube containing EDTA which was stored at 2–8 degrees Celsius and tested within three days. No preparation was required. HbA2/F calibrators and normal and abnormal controls were analyzed at the beginning of each run. Patients with incomplete data were excluded from this study. We analyzed for age, sex variation with Variant of Hemoglobin. A total of 783patients data were evaluated, and the data were entered into Microsoft Excel.

Results:

A total of 783 hemoglobin electrophoresis reports were analyzed at Dr. Benzamin's Pediatric Liver Research Centre and Nutrition Clinic, Sylhet. Out of these, 291(37.2%) were male, 492 (62.8%) female, pediatric population that is under 18 years 387 (44.4%) and 18 years or more 396(50.6%). Among these, 262 (33.46%) reports showed Hemoglobinopathies. Before 6 months, 10 patients were advised for Hb electrophoresis.

Table 1: Age- and sex-wise distribution of cases in pediatric age group

Category	N (%)
Age (mean) months	77.4±73.04
Sex Male Female	 206 (53.2) 181(46.8)
Normal	203(52.5)
Inconclusive	36(9.3)
Hemoglobinopathies	148(38.2%)

Table 2: Age- and sex-wise distribution of cases in adult age group

Category	N (%)
Age (mean) years	43.20±16.96
Sex Male Female	 85 (21.5) 311 (78.5)
Normal	234(59.1%)
Inconclusive	48(12.1%)
Hemoglobinopathies	114(28.8%)

Overall, in study population, most common hemoglobinopathies were Beta thalassemia trait (118, 15.1%), followed by Hb E trait (45, 11.6%), Hb E Beta thalassemia (34, 4.43%), Beta thalassemia major (8, 0.9%). Same trend followed in pediatric age group, Beta thalassemia trait (53, 13.7%), Hb E tarit (84, 10.7%), Hb E Beta thalassemia(28,7.24%), Beta thalassemia major (8, 1.8 %). In adult age group, Betathalassemia trait (65, 16.4%), Hb E tarit (39, 9.8%), Hb E Beta thalassemia (6, 1.5%%).

Table 3: Spectrum of hemoglobinopathies

Category	Pediatric age group n= 387(%)	Adult age group n= 396(%)	Total n=783(%)
Beta thalassemia trait	53(13.7)	65(16.4)	118 (15.1)
Beta halassemia Major	8(1.8)	0	8 (0.9)
Hb E Beta thalassemia	28(7.24)	6(1.5%)	34 (4.43)
HB E disease	2(0.5)	2(0.5%)	4 (0.5)
Hb E trait	45(11.6)	39(9.8%)	84 (10.7)
Alpha thalassemia trait	2(0.5)	0	2 (0.3)
HB d trait	1(0.3)	0	1 (0.1)
Hb D beta thalassaemia	2(0.5)	0	2 (0.3)
Sickle cell Beta Thalassemia	2(0.5)	0	2 (0.3)
sickle cell trait	3(0.8)	2(0.5)	5 (0.6)
sickle cell disease	1 (0.3)	0	1 (0.1)
Hb S/E disease	1(0.3)	0	1 (0.1)

Discussion:

Hemoglobinopathies are the most common inherited red cell disorders worldwide. These diseases create medical, social and economic burdens to the family and to the public.[2]Health-related quality of life and family functioning in parents of children with thalassemia are lower score in all domains of HRQOL and family relationship.[6]

In our study, Hb abnormalities were detected in 262 (33.46%) cases which denotes that disease of Hb disorders is a very significant genetic problem in Bangladesh. Multiple studies found that around 3% and 4% of Bangladeshi population are carrier of β Thalassemia and Hb E respectively.[4] But this scenario is different among the tribal (Chakma, Garo and Marma) populations of Bangladesh, where about 62% population have abnormal Hemoglobin variants with 4.8% Beta thalassemia trait, 35.6% Hemoglobin E trait, 22% Hemoglobin E disease. Cumulative percentage of beta thalassemia and Hemoglobin E trait and E disease were 62.4 %.[7]

A community-based study in Sylhet, Bangladesh, identified at least one inherited blood disorder in 11% of women and 10% of children. Alpha thalassemia was most prevalent, identified in 7% of women and 5% of children, followed by beta thalassemia and hemoglobin E in 2–3%.[8] Our study also found, Beta thalassemia trait (15.1%), Hb E tarit (11.6) %), Hb E Beta thalassemia (4.43%) as common hemoglobinopathies among Sylhet's anemic population. Another study in Bangladesh done by Uddin MM et al., had almost similar findings, that is β-Thalassemia trait (21.3%), E-β-Thalassemia (13.5%)and HbE trait (12.1%) , HbE disease (9.2%) and β-Thalassemia major (0.5%). The majority of the hemoglobinopathies belonged to childhood period (1–15 years), followed by reproductive age group (16–45 years). Few old-age (46+ years) cases were also detected in course of clinical complications.[9] In our study, earliest diagnosis was done at 6 months (2 β-thalassemia major and 1 Hb E disease) and oldest patient was 80 years having Beta thalassemia trait. In pediatric age group, we found Beta thalassemia trait (13.7%), Hb E tarit (10.7%), Hb E Beta thalassemia (7.24%), Beta thalassemia major (1.8)%), sickle/beta thalassemia (0.5%), sickle cell disease (0.3%). Study done by Khan WA et al. at Shishu (Children) Hospital, Dhaka had similar findings with β thalassemia trait (17.94%), HbE β-thalassemia (10.87%) followed by β thalassemia major (4.00%), Hb E trait (12.50%), Hb E disease (2.05%) and Hb D trait (0.35%).[1] Almost similar findings also observed in study done in different region of Bangladesh.[10,11,12] But our findings is different from the study done on Pakistan, they showed beta-thalassemia major (29.4%), followed by sickle / beta-thalassemia compound heterozygous (28.3%), sickle cell disease (28%), Alphathalassemia" (2.8%), HbD trait (2.3%), HbE/Beta-thalassemia(2%), HbD Homozygous (1.8%), Hb D/betathalassemia (1.5%), HbE trait (1.1%) and Beta-thalassemia minor (0.9%).[2] Beta thalassemia trait is also common Hemoglobinopathies in India.[13] But scenario in Europe and America is totally different. They have very low incidence and prevalence of Thalassemia.[14,15]

Conclusion

Hemoglobinopathies among the anaemic population of Sylhet is very common. Beta thalassemia trait, Hb E tarit, Hb E Beta thalassemia, Beta thalassemia major are common variants. So, during anaemia evaluation, hemoglobinopathies should be kept in mind.

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