Review Article

Electrophoretic Pattern of Hereditary Haemoglobin Disorders In Bangladesh.

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Abstract:

Background: Genitic defects of haemoglobin are the most common genetic disorders and affect around 7% of world Population, occur in tropical and sub tropical areas. Beta-thalaraemia is more common in the Mediterranean region while Alfa -thabssaemia is more common in the Far-East.

Objective: To Find out the Pattern of haemoglobin disorders. To evaluate and compare the diseases in study Population. Methods: For this, a total number of 210 subjects with age ranged from 2 to 72 years of both sexes are included in the study. The present study was conducted in out patient department (OPD) of Haematolegy, Bangobondhu Shaikh Muzib Medical University (BSMMU) Dhaka. Bangladesh. During the period of January 2002 to December 2002, Patient ware selected on the basis of morphological blood film examination and Hb-electrophoresis on cellulose acetate at PH 8.6.

Result: Among the 210 Subjects, thalassaemia Trait were (47.14%), Hb-E-beta thalassamia ware 30.47%, Hb E Trait 13.3%, Hb E disease (5.71%) and thalasscmia major ware (3.33%).

Conclusions: It is evident that, Hereditary Haemoglobin disorders are quite common in Bangladesh and this disorders are in herited as autosomal dominant mendalian pattern affecting both male and female.

Key wards: Electrophorctic Pattern; Haemoglobin disorders.

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Introduction:

Hereditary haemoglobin disorders are a heterogeneous group of mandelion disorders. It includes haemoglonopathies are characterized by structurally abnormal haemoglobin variants and thalassemia by partial or total suppression of normal peptide chains of haemoglobin molecules.[1]

More than hundreds of structural haemoglobin variants have been identified in the last three decades. Majority of these results from single aminoacid substitution in one or other of the globing chains. The simple system of presumptive identification of these variants by simple electrophoresis still remain an extremely useful procedure through it does not discriminate between different mutants which carry the same electrophoresis.[2,3,4]

The inheritance of haemoglobin disorders follows a simple mandelian pattern. The heterozygous state for a disorder is called "Trait" while the homozygous or genetic compound is called "disease". Thalasseamia is most common inherited genetic disorder and varies in different population group in the world. Haemoglobin disorder will become a major issue in developing countries like Bangladesh in this millennium. World Health Organization (WHO) estimates that at least 7% of the world population carriers of different inherited disorder of haemoglobin. It is observed that when the world population finally stabilizes at least 8% of the population will be carrier or Trait[5]. The world population of carrier of beta thalassaemia trait is reported to be more than 100 millions world wide and about 1,00,000 children with thalassaemia major are born each year.

In Banglsdesh, there is no definite data regarding electrophoresis pattern of hereditary haemoglobin disorders. No screening program has been taken any population group. A conservative world health report estimates that 3% are carrier of Beta thalasseamia and 4% are carriers of Hb E in Bangladesh[7].

Most of the thalassaemic patients need frequent blood transfusion about every 2-3 weeks interval. As a result good percentage of blood is utilized by them, which major burden to the department of transfusion medicine with lot of complication and transfusion hazards. So there is maximum chance of transmission of infectious agent like HCV, HBV, HIV, plasmodium & treponema pallelum etc.

Methods:

The present study was conducted in Out Patient Department (OPD) of Haematology Department, Bangobondhu Shaikh Mozib Medical University (BSMMU), Dhaka. Out Patient Department (OPD) of Haematology could provide adequate number of cases for this study. The study was carried out from January to December, 2002 and total No. of 210 cases of hereditary haemoglobin disorder were studied over a period of one year. As Dhaka in the capital city and received the patient which referred from all parts of the country. It seems that the sample roughly represents the whole population of country.

Patient were selected on the basis of morphological evidence of heamolytic anaemia in peripheral blood film and haemoglobin electrophoresis on cellulose in stale at PH 8.6, Age, Sex Presenting complaints and family history were noted Table I, II, III & IV, Diagram.

Result:

Among the 210 Subjects, thalassaemia Trait were (47.14%), Hb-E-beta thalassamia ware 30.47%, Hb E Trait 13.3%, Hb E disease (5.71%) and thalasscmia major ware (3.33%).

Table – 1 : Distribution of population by age

Age (years)	No. of patient	Percentage %
<5	35	16.66%
5-10	32	15.23%
11-20	60	28.57%
21-30	56	26.66%
>30	27	12.88%

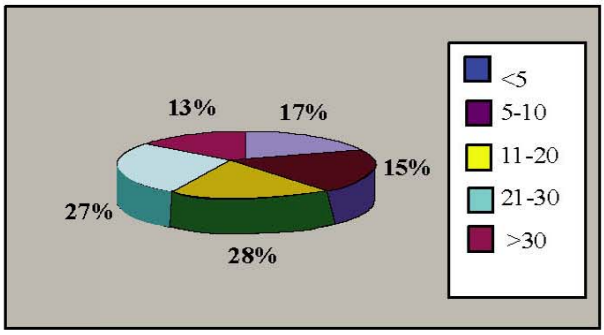

Fig : 1- Distribution of population by age

Table – ll : Sex distribution

Sex	No. of patient	Percentage %
Male	108	51.43%
Female	102	48.57%
Total	210	100%

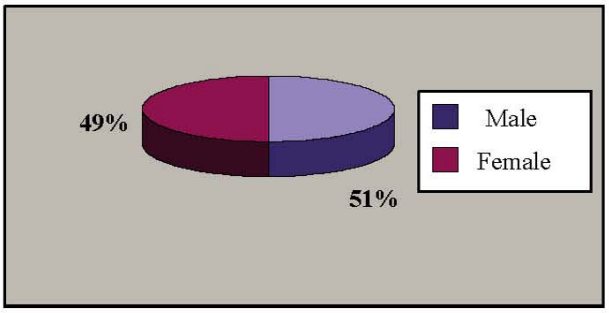

Fig : 2- Sex distribution

Table – lll : Presenting clinical manifestation

Symptoms & sign	No. of patient	Percentage %
Weakness	148	70.47%
Jaundice	117	55.71%
Hepatomegaly	84	40.00%
Splenomegaly	143	60.09%
Fever	76	36.19%
Retardation of growth	27	12.89%
Pallor	6	2.85%
Leg ulcer	5	2.38%
Bony cange	4	1.48%
Asymptomatic	53	25.23%

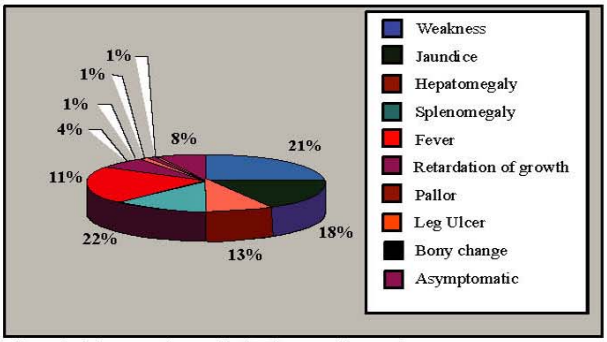

Fig : 3- Presenting clinical manifestation

Table – 1V: Pattern of hereditary haemoglobin disorders.

Symptoms & sign	No. of patient	Percentage %
Beta-thalassacmia trait	99	47.14%
Hb- E-beta thalassemia	65	30.47%
Hb-E-Trait	28	13.33%
Hb-E disease	12	5.71%
Beta-thalassemia Major	7	3.33%

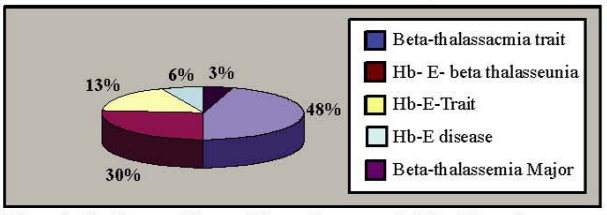

Fig : 4- Pattern of hereditary haemoglobin disorders.

Discussion:

In this study heterozygous beta thalassaemia was foud most common. Double heterozygous Hb-E-beta thalassemia and heterozygous HbE, were next more common. Similar findings observed in the others studies.[8,9]

In our country, data regarding the hereditary haemoglobin disorder are not available so far, but in our neighboring countries in Myanmar and India, they have got their prevalence rate. Bangladesh is in geographical continuity with Myanmar, Assam (India), West Bengal (India), Tripura and also same belt of Thailand and Combodia. And also the population of West Bengal, Assam and Myanmar share the same ancestry with that of Bangladesh. In North Eastern, the Hb-E gene reaches the frequencies of about 7.5%[10], Myanmar it is about 10-20% and Assam it is 30%. As Bangladesh is situated in Between all these area and the people might have been migrated from these areas to Bangladesh in decades earlier.

In this study observed that the prevalence rate of HB-E gene (13.33%|) is quite similar to other neighboring countries. In one study in West Bengal (Calculla) showed that the prevalence of beta thalassaemia trait alone to be 7.5%. In West Bengal and it is much higher 12.6% in Orissa (India)[11]. It can compare this observation regarding the incidence also correlate well with some of the small studies done Bangladesh on hereditary haemolytic anaemia[12]. But incidence as well as percentage study is varied because this present study was done only from pattern of electrophoresis in cellulose agar acetate at PH 8.6. Within this small study group Hb-E beta thalassaemia and Hb-E trait have taken a place. It is observed from other studies double heterozygous Hb-E beta thalassaemia was the commonest thalassaemia syndrome[12,13]. Disorders are manifested at all ages from minimum 1.5 years to 72 years. The highest incidence in second decade (28.37%). The age distribution shown in the table roughly correlate by other study.[13]

Conclusion:

It is evident from this study, the Hereditary haemoglobin disorders are quite common in Bangladesh and these disorders are inherited as autosomal dominant mendelion pattern affecting both male & female. So we can not avoid these diseases. In this study, we got heterozygous (trait) like heterozygous both thalassaemia and heterozygous Hb-E trait significant number in camouflage. These population are usually asymptomatic, do not require treatment and lead a reasonably good quality of life but they are dangerous because of possibility of homozygous or double heterozygous inheritance through marriage of unaware couples or silent spread as trait. This is a serious health threat to our nation if it is allowed to continue without taking measures for prevention.

Finally in spite of all limitations of the study we have at least reached a concrete conclusion that hereditary haemoglobin disorders is a very common problem in Bangladesh, on which the health authorities should focus. An awareness has to be created at the national level to reduce the incidence of hereditary haemoglobin disorders in community. It is mandatory to detect the trait in general population with large scale and proper genetic counseling should be ensured. It is the time to think about the molecular and prenatal diagnosis to start to prevent the further spread of the diseases.

Aclcnowledgement:

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