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Prevalence of Thalassaemia and Other Hemoglobinopathies In A Northern District Of West Bengal, India

Dr. Subhrajyoti Naskar [1] , Dr. Gouranga Biswas [2] ,Dr.Sudip Ranjan Paul [3] ,Dr. Sujit Das [4] ,Dr. Debasis Das[5] , Dr. Indranil Dawn[6] , Dr. Apu Adhikari [7]

[1,] *Assistant Professor, Deptt. of Community Medicine, Malda Medical College, Malda.*
[2,] *Assistant professor, Deptt. of Pediatrics, Malda Medical College, Malda.*
[3,] *RMO, Deptt. of Gynaecology and Obstetics, Malda Medical College, Malda.*
[4,] *Medical Officer , Thalassemia unit, Malda Malda Medical College, Malda.*
[5,] *Associate Professor, Community Medicine, Malda Medical College, Malda*
[6,] *Assistant Professor, Deptt. of Biochemistry, Malda Medical College, Malda.*
[7,] *R.M.O. Deptt. of Medicine North Bengal Medical College, siliguri*

Abstract: Background: *Hemoglobinopathies are disorders affecting the structure, function, or production of hemoglobin. It is estimated that 8000-10000 children are born with thalassemia major every year in India. There are around 65000 thalassemia patients in our country at any given time.*

Objectives: *To assess the prevalence of thalassaemia and other hemoglobinopathies in a northern district of West Bengal, India.*

Material And Methods: *The Study was conducted at Malda Medical College, West Bengal – a rural tertiary care Health Care Institution during the period of january2012 to January 2013. High-performance liquid chromatography (HPLC), complete blood count (CBC) and hemagglutination technique were performed for the assessment of abnormal hemoglobin variants .*

Result: *Among 5156 total all types of population surveyed 12.88% were found to be any type of thalassaemia carrier and prevalence of all sorts of Thalassaemia were found 2.68% ;3.04%, HbE carrier – 9.02%, HbS carrier – 0.35% and 12.88% other carriers. Among 1819 antenatal mothers0.44% affected found; Among 65 children, 36.92% total Thalassaemic found; Among 2971 premarital population surveyed, 3.13% Thalassaemic were fond; Among 301 post-marital population, 4.31% affected member found.****Conclusion:*** *High prevalence of hemoglobinopathies where Beta thalassaemia in heterozygous stated occurred more frequent than other hemoglobinopathies.*

Keywords: *Thalassaemia, prevalence, Malda district.*

I. Introduction

Hemoglobin is critical for normal oxygen delivery to tissues; it is also present in erythrocytes in such high concentration that it can alter red cell shape, deformability, and viscosity. [1]

Hemoglobinopathies are disorders affecting the structure, function, or production of hemoglobin.

This conditions are usually inherited and range in severity from asymptomatic laboratory abnormalities to death in utero. Different forms may present as hemolytic anemia, erythrocytosis, cyanosis, or vasoocclusive stigmata. (4),2 There are five major classes of hemoglobinopathies:

1.Thalassemia Syndromes

Thalassemias are genetically transmitted disorders. Normally, an individual inherits two beta- globin genes located one each on two chromosomes 11, two alfa- globin genes one each on two chromosomes 16, from each parent normal adult hemoglobin(HBA) is A2B2. Depending upon whether the genetic defect or deletion lies in transmission of alfa or beta- globin chain genes. Thalassemias are classified into alfa-thalassemia, beta- thalassemia and Delta beta, gama delta beta, alfa beta thalassemia. (4)

2- Structural Hemoglobinopathies-

Structural Hemoglobinopathies occours when mutations alter the amino acid sequence of a globin chain, altering the physiologic properties of the variant hemoglobins and producing the characteristic clinical abnormalities. The most clinically relevant variant hemoglobins polymerize abnormally, as in sickle cell anemia, or exhibit alted solubility or oxygen- binding affinity . (4) Most common hemoglobinopathie is sickle cell syndrome.(3)

3-Thalassemic Hemoglobin Variants- structurally abnormal Hb associated with co- inherited thalassemia phenotype
One of the important variant is HbE

4-Hereditary Persistence Of Fetal Hemoglobin
5-Acquired Hemoglobinopathies

Hemoglobinopathies are especially in areas in wich malaria is endemic suggesting that nature developed genetic mutation to overcome mortality and morbidity of malaria.Thalalassemia are the most common genetic disorders in the world, affecting nearly 200 million people worldwide. About 15% of American blacks are silent carriers for alfa-thalassemia; alfa- thalassemia trait(minor) occours in 3% and in 1-15% of patient in persons of Mediterranean origin. Beta- thalassemia has a 10-15% incidence in individuals from the Mediterranean and South-East Asia and 0.8% in American blacks.(4)

There are >200 muations for Beta- thalassemia, although most are rare. About 20 common alles constitute 80% of the known thalassemias worldwide; 3% of the world's population carry genes for beta-thalassemia.(1) Thalassemia incidence varies in various communities, religions and ethnic groups in India. Ahigher frequency is noted in certain communities such as in Sindhis and Punjabis .2 In india, prevalence of the beta-gene varies from 1%-17%. It is estimated that 8000-10000 children are born with thalassemia major every year in India. There are around 65000-67000 thalassemia patients in our country, at any given time.

With the above perspectives the present study was carried out to assess the Prevalence of thalassaemia and other hemoglobinopathies in a northern district of West Bengal, India.

II. Material And Methods

The Study was conducted at Malda Medical College, West Bengal – a rural tertiary care Health Care Institution during the period of january2012 to January 2013. All the patients attending Thalassaemia Clinic and all the outreach camp conducted by the clinic during the period of study constituted the study population. No sampling was done; census method adopted. High-performance liquid chromatography (HPLC), complete blood count (CBC) and hemagglutination technique were performed for the assessment of abnormal hemoglobin variants.

III. Result

The general population has been divided under four group i.e., Antenatal mother, Children, premarital group of population and post-marital population. Prevalence were calculated for each sub-group for Beta thalassaemia carrier, HbE carrier, HbS carrier and other carrier which includes HbE traits and HbD traits.

Among 1819 general antenatal mother studied total carrier 177(9.73%). Among these carrier 55(3.02%) were B Thalassaemia carrier, 110(6.05%) HbE carrier, 6(0.33%) Hbs Carrier and another 6(0.33%) were other carrier. Among 65 children studied, 1(1.54%) were B thalassaemia carrier, 5(7.69%) HbE carrier; total carrier being 6(9.23%). Among 2971 premarital population, 421(14.17%) were carrier; 82(2.76%) B Thalassaemia Carrier, 321(10.5%) HbE carrier, 10(0.34%) HbS carrer and 17(0.57%) other carrier. 301 Post-marital population were surveyed among which 60(19.93) were carrier, 19(6.31%) being B Thalassaemia carrier 38(12.62%) were HbE carrier, 2(0.66%) HbS carrier and 1(0.33%) other carrier. Among 5156 total all types of population surveyed 664(12.88%) were found to be any type of thalassaemia carrier; prevalence of B Thalassaemia carrier being 157(3.04%), HbE carrier – 465(9.02%), HbS carrier – 18(0.35%) and 664(12.88) other carriers. (Table 1) For assessing prevalence of at risk population 320 family members of diseased and suspects and suspected patients were tested. Among 299 family members of diseased/ suspected persons, 157(52.51%) were found carrier of all types of Thalassaemia; 80(26.76%) – B Thalassaemia carrier, 74(24.75%) HbE carrier, 2(0.67%) – HbS carrier and 1(0.33%) were other carrier. Among suspected patient, 2(9.52%) were found to B Thalassaemia carrier; none other types of carrier found in these group. (Table 2)

Among 5156 general population studied, prevalence of all sorts of Thalassaemia were found 138(2.68%). Among 1819 antenatal mothers 8(0.44%) affected found; 1(0.06%) – HbE beta thalassaemia, 6(0.33%) – HbE homozygous and 1(0.06%) were others. Among 65 children, 24(36.92%) total Thalassaemic found; 1(3.08%) being B Thalassaemic, 21(32.31%) HbE beta Thalassaemic, and 1(1.54%) HbE homozygous Thalassaemic. Among 2971 premarital population surveyed, 93(3.13%) Thalassaemic were fond; 8(0.27%) being HbE beta Thalassaemic, 83((2.79%) – HbE homozygous and 2(0.07%) – others. Among 301 post-marital population, 13(4.31%) affected member found; 2(0.66%) being HbE beta thalassemia and 11(3.65%) - HbE homozygous. (Table 3) Among 320 at risk population, prevalence of all types of Thalassaemia were 52(16.25%); 4(1.25%)– B Thalassaemia, 35(10.94%) - HbE beta thalassemia, 13(4.06%) being Hb Homozygous. Among 299 family members of affected person, 40(13.38%) were affected; 3(1.00%) – B Thalassaemia, 24(8.03%) – HbE beta Thalassaemia and 13(4.35%) – HbE homozygous. Among 21 suspected patient 12(57.14%) were affected; 1(4.76%) – B Thalassaemia and 11(52.38%) BetaThalassaemia. (Table 4)

IV. Discussion

Study conducted by Mondal B et al at B.S.Medical College, West Bengal, another neighouring district rural medical college taking 958 patiets over 6 months of 2011 at the hospital clinic showed high prevalence of hemoglobinopathies (27.35%) where Beta thalassaemia in heterozygous stated occurred more frequent than other hemoglobinopathies. Out of 958 patients, 72.65% were HbAA and 27.35% were hemoglobinopathies individuals where 17.64% β-thalassemia heterozygous, 2.92% β-thalassemia homozygous, 3.86% HbAE, 1.15% HbAS trait, 1.25% HbE-β thalassemia trait and 0.52% HbS- Beta thalassemia trait were found.[2] Study of Jain BB et al at Burdwan Medical College and Hospital - another neighbouring district rural medical college showed the importance of hospital based screening of population in absence of community based diagnosis register. In their study they studied prevalence of all hemoglobinopathies over 3 years 4 months which was 29.3% among hospital clinic attendant. In their study also Beta thalassaemia heterozygous was the most common hemoglobinopathy in that area closely followed by hemoglobin E heterozygous. In their study no outreach screening was there and they advocated a routine premarital screening program for identification and prevention of high-risk marriages.[3]

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Tables:

Table 1. Prevalence of thalassaemia carrier among general population under study.

General population	B thal carrier No.(%)	HbE carrier No.(%)	HbS carrier No.(%)	Other carrier(HbE trait, HbD)No.(%)	Total carrier No.(%)	Total population No.(%)
Antenatal	55(3.02)	110(6.05)	6(0.33)	6(0.33)	177(9.73)	1819(100.00)
Children	1(1.54)	5(7.69)	0(0.00)	0(0.00)	6(9.23)	65(100.00)
Premarital	82(2.76)	312(10.50)	10(0.34)	17(0.57)	421(14.17)	2971(100.00)
Post-marital	19(6.31)	38(12.62)	2(0.66)	1(0.33)	60(19.93)	301(100.00)
Total	157(3.04)	465(9.02)	18(0.35)	24(0.47)	664(12.88)	5156(100.00)

Hb E carrier is more in total population. Then B Thal carrier. Among antenatal mother it is also high.

Table 2. Prevalence of thalassaemia carrier among at risk population under study.

At risk population	B thal carrier No.(%)	HbE carrier No.(%)	HbS carrier No.(%)	Other carrier (HbE trait, HbD)No.(%)	Total No.(%)	Total population No.(%)
Family of diseased/suspects	80(26.76)	74(24.75)	2(0.67)	1(0.33)	157(52.51)	299(100.00)
Suspected patient	2(9.52)	0(0.00)	0(0.00)	0(0.00)	2(9.52)	21(100.00)
Total	82(25.63)	74(23.13)	2(0.63)	1(0.31)	159(49.69)	320(100.00)

Table 3. Variation of Talassaemia among general population under study.

General population	B thalassaemia No.(%)	HbE beta thalassemia No.(%)	HbE homozygous No.(%)	HbS homozygous No.(%)	Others (HbE heterozygous) No.(%)	Total No.(%)	Total population No.(%)
Antenatal	0(0.00)	1(0.06)	6(0.33)	0(0.00)	1(0.06)	8(0.44)	1819(100.00)
Children	2(3.08)	21(32.31)	1(1.54)	0(0.00)	0(0.00)	24(36.92)	65(100.00)
Premarital	0(0.00)	8(0.27)	83(2.79)	0(0.00)	2(0.07)	93(3.13)	2971(100.00)
Post-marital	0(0.00)	2(0.66)	11(3.65)	0(0.00)	0(0.00)	13(4.31)	301(100.00)
Total	2(0.04)	32(0.62)	101(1.96)	0(0.00)	3(0.06)	138(2.68)	5156(100.00)

Table 4. . Variation of Talassaemia among at risk population under study.

At risk population	B thal No.(%)	HbE beta thalassemiaNo.(%)	HbE homozygous No.(%)	Total No.(%)	Total population No.(%)
Family of diseased/suspects	3(1.00)	24(8.03)	13(4.35)	40(13.38)	299(100.00)
Suspected patient	1(4.76)	11(52.38)	0(0.00)	12(57.14)	21(100.00)
Total	4(1.25)	35(10.94)	13(4.06)	52(16.25)	320(100.00)