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

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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 January 2012 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 mothers 0.44% affected found; Among 65 children, 36.92% total Thalassaemic found; Among 2971 premarital population surveyed, 3.13% Thalassaemic were found; 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. 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)

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 alpha- 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 altered solubility or oxygen- binding affinity. (4) Most common hemoglobinopathy 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

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 carrier 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 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 type 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 Thalassaemia 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%) Thalassaemia were found; 8(0.27%) being HbE beta Thalassaemia, 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 thalassaemia 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 thalassaemia, 13(4.06%) being Hb Homozygous. Among 299 family members of affected person, 40(13.8%) 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)

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IV. Discussion

Study conducted by Mondal B et al at B.S.Medical College, West Bengal, another neighbouring district rural medical college taking 958 patients 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. 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 premartial screening program for identification and prevention of high-risk marriages. 

References