

Study on Pattern of Chronic Haemolytic Anemia among the Tribal and Nontribal Paediatric Population of Rural Bengal

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Abstract: Chronic haemolytic anemia occurs due to haemolysis of RBC. It may be due to defect of haemoglobin synthesis and structure, or due to abnormal RBC structure. It is a major debilitating disease among the paediatric population often requiring repeated blood transfusion. As most of the chronic haemolytic anemia are genetically transmitted, the condition is worst in the tribal areas where rate of consanguineous marriage is quite high. This is reflected in different studies which show high prevalence of sickle cell disease and G-6-P-D among tribal population. However no reported study on the complete profile of chronic haemolytic anemia among the tribal paediatric population has not been found. So the present study conducted in a tertiary care hospital in Bankura district of west Bengal, centering most of the tribal population of west Bengal has been conducted to find out the pattern of chronic haemolytic anemia among the tribal and nontribal paediatric population. Over a study period of 1 year about 120 chronic haemolytic patients were registered. Out of 120 patients 80 belonged to nontribal population and 40 were tribal. Out of 120 cases 50 were thalassemia major accounting for 41.6% of the chronic haemolytic anemia was the most common etiology, followed by E-beta thalassemia (35%). Sickle cell disease, G-6-P-D, AIHA (autoimmune haemolytic anemia) were particularly common among the tribes. The average age of presentation being 2.09 years.

Keyword: Chronic haemolytic anemia (CHA), β -thalassemia major, E- β thalassemia, Sickle cell disease and G6Pd deficiency, Tribal.

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I. Introduction

Chronic haemolytic anemia refers to haemolytic anemia which is primarily due to increased haemolysis of RBCs. The defect may be due to abnormal haemoglobin structure or due to abnormal RBC structure. Increased RBC destruction may be due to

1) Intrinsic abnormality of RBC-

a) Hereditary red cell enzyme defect eg-G-6-P-D, pyruvate kinase deficiency.

b) Hereditary red cell membrane defect-hereditary spherocytosis.

2) Ineffective erythropoiesis due to haemoglobinopathies—thalassemia syndrome

3) Abnormal haemoglobin- sickle cell anemia

4) Autoimmune haemolytic anemia

5) Acquired- drugs, malaria

The first study that threw light on haemoglobinopathies in west Bengal by Chatterje¹ in 1959 showed the presence of 87% cases of HbE Thalassemia, the rest comprising mainly of homozygous thalassemia. While HbE thalassemia is common in West Bengal and Bangladesh, there are many studies showing increased prevalence of sickle cell anemia among tribes. However there has not been any study revealing the complete profile of chronic haemolytic anemia among paediatric population and among the tribal population. This study is an attempt to study the pattern of chronic haemolytic anemia among the paediatric population of rural Bengal with special emphasis on comparison of the pattern of chronic haemolytic anemia between the tribals and the nontribals.

II. Material And Methods

The present descriptive Observational study has been done to find out the pattern of distribution of chronic haemolytic anemia among tribals and nontribals.

The present study recruited 120 cases diagnosed as chronic haemolytic anemia within the study period of 1 year. Sample was selected from general paediatric population attending OPD or indoor admission in paediatric ward.

Children aged 6months to 8 years of both sexes irrespective of caste and socioeconomic status who presented with pallor for more than six months along with any of the features of chronic haemolytic anemia like:

a)Hepatosplenomegaly b) corrected reticulocyte count >3% c) evidence of haemolysis in peripheral blood smear like target cells, normoblasts, spherocytes, fragmented cells atc were included in the study.

Any child with pallor <6months, corrected reticulocyte count <3%,peripheral blood smear without evidence of haemolysis in peripheral blood smear were excluded from the study.

After proper education all parents of the cases gave their written consent to allow inclusion of their child in the study. A predesigned proforma was filled for every case which included a detailed history including age, sex, ethnic origin,systemic examination and laboratory investigations.The laboratory investigations done for the study per se were as follows- complete blood count with reticulocyte count and peripheral blood smear,osmotic fragility test, sickling test, G-6-P-D assay,serum ferritin assay, and direct comb test. All the statistical analyses were done according to the SPSS software.Approval was obtained from the institutional ethical committee before starting the study.

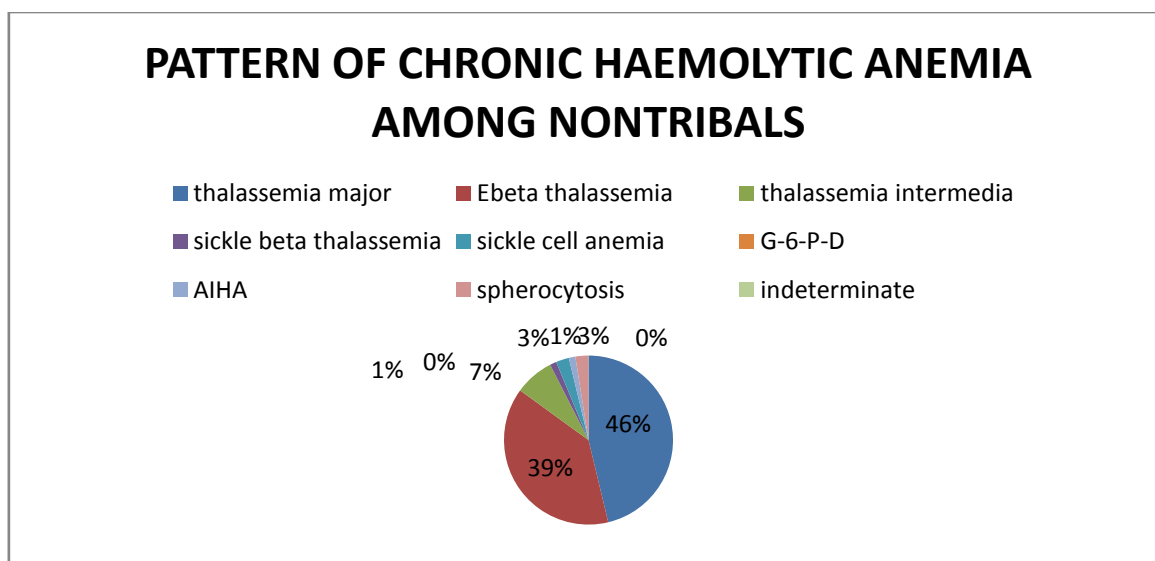
III. Result

In the present study,120 cases of chronic haemolytic anemia which consists of both male and female of both tribe and nontribal community were included.

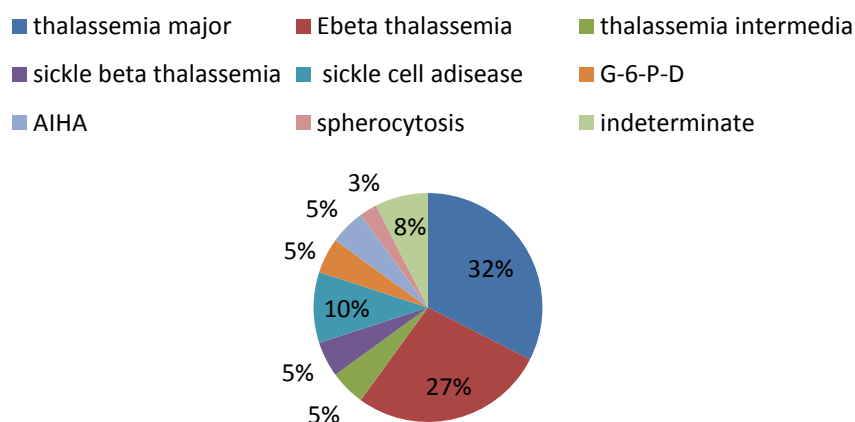
Among the chronic haemolytic anemia, thalassemia syndrome occupied the major portion i.e. 103/120 cases(85.8%) followed by sickle cell disease 6/120(5%).other chronic haemolytic anemia diagnosed were G-6-P-D 2/120(1.6%),Autoimmune hemolytic anemia 3/120(2.5%), spherocytosis 3/120(2.5%) respectively. In spite of all the investigations 3out of 120 cases etiology were undiagnosed and hence classified as indeterminate CHA.Thalassemia major accounted for 50/120(41.6%) followed by Ebeta thalassemia 42/120(35%).Thalassemia intermedia was found among 6.6%,Sickle beta thalassemia was 2.5%.

74/120(61.6%)caseswere male and 46/120 (38.3%) were females. Male female ratio of beta thalassemia major, E beta thalassemia,thalassemia intermedia and sickle beta were as follows 3:2, 4:3,3:1 and 2:1 respectively. Male female ratio of sickle cell disease and spherocytosis was 5:1 and 1:2 respectively. Only 2 cases of G-6-P-D were found and both of them were males.

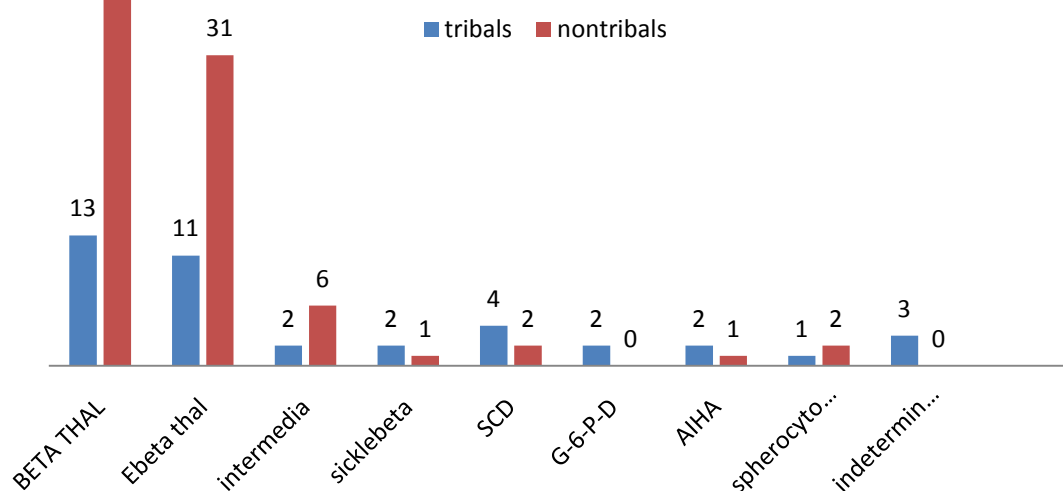
The mean age of presentation of the entire chronic haemolytic anemia in our study was 2.6years. for studying the age distribution 4 age groups were taken 6month -1 year,>1year-2 years,>2years – 5years,>5years -8years. It was found that 26/50(52%) cases of thalassemia major were diagnosed at the age 1-2years with mean age of presentation being 1.3years. In case of Ebeta thalassemia was 2.09years with 52% in age group 1-2years and 35% in 2-5years. Both in beta thalassemia major and E beta thalassemia no cases were detected after 5 years. This could be attributed due to the fatal outcome of the undiagnosed cases. Thalassemia intermedia 62.5% were detected in age group 3.2years. Sickle beta cell disease and sickle cell disease the mean age was 2.2years and 2.02 years respectively. AIHA,spherocytosis on the other hand had the mean age of presentation 5.28 years and 5.42years respectively. 2 cases of G-6-P-D were diagnosed having the mean age of diagnosis 4 years. 3 indetermined CHA were there with mean age of diagnosis 1.1years. Mean age detection of CHA among tribals was 2.5years and that among nontribals was 1.4years. Girls with CHA were detected at a mean age of 2years and that among boys was 1.3years. This was found to be statistically significant(p=.002) this can be due to the fact less awareness among the tribals,difficult access to health facilities and negligence regarding girl child.



PATTERN OF CHRONIC HAEMOLYTIC ANEMIA AMONG TRIBALS



COMPARISION OF DIFFERENT TYPES OF CHRONIC HAEMOLYTIC ANEMIA AMONG TRIBALS AND NONTRIBALS



Among 120 cases 40 were tribal and 80 were nontribal. Male female ratio among tribals 1.5:1 and that among nontribals was 1.6:1. Among beta thalassemia major 13/50(26%) were tribal and 37/50(74%) were nontribal. Among Ebeta thalassemia 11/42(26.1%) were tribal and 31/42(73%) were nontribal. Thalassemia intermedia 2/8(25%) were tribal and 6/8(75%) were nontribal. Among sickle cell disease 4/6 (66.6%) were tribal and 2/6(33.3%) were nontribal. Sickle beta thalassemia 2/3(66.6%) were tribal and 1/3(33.3%) were nontribal. AIHA 2/3(66.6%) were tribal and 1/3 (33.3%) were nontribal. This clearly shows tribal population have higher percentage of sickle cell anemia and AIHA which is statistically significant ($p=0.003$). All the cases of G-6-P-D were of tribal origin. 3 cases of indetermined etiology were found among the tribals, signifying the possibility of presence of a type of CHA yet to be discovered for which further studies on large scale on tribal population over a longer period of time is needed.

IV. Discussion

Though exact incidence and prevalence can only be estimated from community study, we have tried to estimate the disease burden of congenital haemolytic anemia in a hospital set up of rural Bengal. However this hospital based study is important for sensitization and can serve as a baseline for generating more data.

Among the chronic haemolytic anemia, Thalassemia syndrome is the commonest in this area. Beta thalassemia was 42% followed by E beta 35%, Thalassemia intermedia was 6.6%, sickle beta thalassemia was 2.5%.

Spectrum of haemolytic anemia in Punjab North India by Dash S & Dash RJ² also showed that thalassemia was the leading cause (40%-50%).

Ambekar, et al. study³ in Western Maharashtra studied for diagnosis of haemoglobinopathies on total of 1291 subjects which included 891 case from pediatric age group and 400 cases from adult population. Their study revealed that among 890 pediatric subjects studied 790 were detected to be normal, 101 showed the presence of one or other haemoglobinopathy. Thalassemia major was detected in 76 (8.5%) subjects, beta thal trait in 4 (.5%), sickle cell disease in 16 (1.8%) HbE in 4 (.5%) and HbD disease in 1 (.1%).

Study of Basu, et al. in and around Kolkata⁴ shows normal HPLC 76.24% and among HPLC asymptomatic cases was 16.33% ; out of which 11.19% was Beta Thalassemia carrier, 4.27% was HbE trait, 0.10% was HbE (homozygous), 0.05% was sickle cell carrier, 0.32% was alpha-thal carrier, 0.2% was Hb-Lepore carrier, 0.01% was Hb-D carrier and 0.10% HPFH. Among symptomatic cases 1% was Beta-thal major, E-beta 2.85% and 0.16% was sickle beta thal.

The study conducted⁵ by Sahana et al. On Congenital Haemolytic Anemia in Bangladesh showed the sex ratio of cases was M: F = 65:61. Ages ranged from 14 months to 12 years with a mean age of 7.5 years. Thirty per cent patients presented with their initial symptoms between the ages of 2-5 years followed by 28%, 22% and 20% who presented between the ages of 5-8 years, 1-2 years and <1 year, respectively. On the other hand majority of the patients (38%) were diagnosed between 5-8 years followed by 26%, 15%, 13% and 8% diagnosed between the ages of 2-5 years, >8 years, 1-2 years and <1 year, respectively. This study found that 67% of 126 cases had HbE-beta Thalassemia and 29% had Beta Thalassemia major or intermedia. Hemoglobin electrophoresis findings showed that HbE-beta Thalassemia patients had 40% to 80% of HbE, 5%-35% of HbF and 0%-30% of HbA. Thalassemia patients showed two patterns of hemoglobin electrophoresis findings. In one group, 19 (53%) patients had 95 - 98% HbF and 2 - 5% HbA₂. Thalassemia patients included both the beta Thalassemia major and beta-Thalassemia intermedia. For 2.4% patients, we could not reach a confirmed diagnosis. Another 1.6% patients had congenital spherocytosis. Compared to this our study shows a much lower age of presentation, reflecting higher level of consciousness regarding chronic haemolytic anemia and much earlier presentation.

I. Panigrahi, et al. study⁶ showed that Hb-E beta is common haemolytic anemia in South East Asia, with varied prevalence of 7% to 50% in different parts of North East Asia, with varied prevalence of 7% to 50% in different parts of North Eastern region of India and 1% to 2% in West Bengal. Hb-E may not be clinically significant, but interaction of it with beta-thalassemia produces variable clinical presentation. In our study also the percentage of E beta thalassemia is 35% among congenital haemolytic anemia, ranking next to beta thalassemia major.

Kamble M, Chaturvedi P. studied the epidemiology of sickle cell disease⁷ in rural hospital in central India. 1753 admissions were screened out of which 99 (5.7%) were diagnosed as SCD of which, 61 (61.6%) were homozygous (HbSS) whereas 38 (38.4%) were heterozygous (HbAS). SCD was more common among boys with male female ratio being 1.65:1 in HbSS and 1.71:1 in HbAs. In the present study the percentage of sickle cell disease is 5% with male female ratio being 5:1. The mean age of presentation is 2.02 years.

The present study corroborates with different studies in India on chronic haemolytic anemia. But the uniqueness of the study lies in its attempt to define the pattern of chronic haemolytic anemia among the tribal community and compare it with nontribal community.

V. Conclusion

β-thalassemia major is the major CHA followed by E-β thalassemia. Sickle cell disease and G6Pd deficiency was more common among tribals. Age of hospital presentation was higher among tribal girls may be due to less awareness among the tribals, difficult access to health facilities and negligence regarding girl child. CHA of indeterminate etiology needs further evaluation.

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