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Original Research Article

Clinico-epidemiological profile of hemolytic anemia in children from a single center in a tertiary care hospital at eastern India

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Abstract
Background: *The hemolytic anemias are group of disorders characterized by increased destruction of RBC while bone marrow compensating for hemolysis by increased erythropoisis. Hemolytic anemias can be due to a corpuscular defect (mainly congenital) or due to an abnormal hemolytic mechanism (extrinsic abnormality).Among the congenital causes, Hemoglobinopathies and thalassemia constitute a major proportion.*
Methods: *A prospective study was carried out among 135 patients of congenital hemolytic anemia in IMS and SUM Hospital, Bhubaneswar, Odisha. a tertiary care teaching hospital at eastern India for a period of one year. Detailed information on demographic pattern, clinical profile and hematological parameters were assessed.*
Results: *Out of the total 135 patients evaluated, it was found that the most common cause of congenital hemolytic anemia was sickle cell trait (35.5%) followed by sickle cell disease (30.4%) beta Thalassemia trait(17.9%), beta thalassemia major (7.4%). The mean hemoglobin was found to be lowest in beta thalassemia major (6.10mg/dl). The most common presenting symptom was hemolytic facies(53.33%) followed by growth retardation (49.62%). Most patients were residing from the district of Nayagarh (46%)*
Conclusion: *The incidence of sickle cell trait is relatively higher in comparison to other causes of hemoglobinopathies and is a major health problem in eastern area of the country.*
Keywords: *Beta thalassemia, Congenital hemolytic anaemia, Sickle cell trait.*

Introduction

The hallmark of hemolytic anemias is an increase in the rate of red blood cell (RBC) destruction. This process reduces the lifespan of 100-120 days; the RBC survival depends on the rate of hemolytic process, which may be mild to very severe. The destruction of RBC occurs due to either an intracorpuscular (intrinsic) abnormality that renders them more susceptible to the normal mechanisms of red cell destruction or an extracorpuscular (extrinsic) abnormality due to development of an abnormal hemolytic mechanism. The intrinsic defects are mainly congenital. Basic defect may be in any of the three main components of the cell- the membrane, the hemoglobin molecule, and various enzymes concerned with cell metabolism. Acquired hemolysis may result from either an immune or a non immune mechanism. Few most common causes of congenital hemolytic anemia are

hemoglobinopathies, like Sickle cell anemia, α Thalassemia, beta Thalassemia, HbE beta Thalassemia; RBC enzyme deformity in thr form of glucose 6 Phosphate Dehydrogenase Deficiency; RBC membrane defects like Hereditary Spherocytosis[1]. The prevalence of beta thalassemia trait is between 3-17% and is related to consanguinity[2]. Every year, ten thousand children with beta thalassemia major are born in India, which constitutes 10% of the all around the world[3]. HbE thalassemia is seen in north-east regions of India[4].

The treatment available for thalassemia patients are standard blood transfusion, iron chelation and splenectomy in cases with hypersplenism. The corrective treatment like bone marrow transplantation is expensive so alternatively the disease burden can be reduced by population screening, genetic counseling and pre-natal diagnosis[5] .

This study conducted in IMS and SUM hospital, Bhubaneswar, Odisha with the aim to find out the clinical and hematological profile of patients with various types of hemolytic anemia.

Methodology

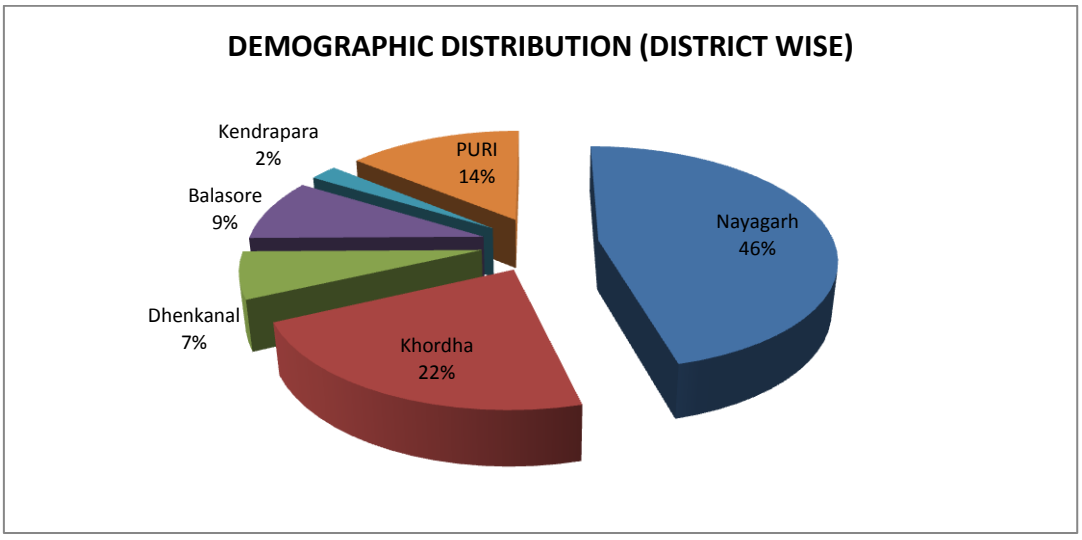
This is a prospective study carried out in a tertiary care teaching Hospital of Odisha over a period of one year from January 2018 to December 2018. All children aged 1 month to 5 years diagnosed with hemoglobinopathies by Hb electrophoresis were included in the study. Patients with serious systemic illness were excluded from the study. The approval of ethical committee was taken prior to the commencement of the study.

Those patient under five years of age attended pediatric IPD or OPD with features of hemolytic anemia, family history of consanguinity, blood transfusion were included in the study. Detailed socio-demographic profile, relevant clinical history, clinical signs and symptoms were collected with the help of pre-validated proforma. Complete blood count (CBC) and Reticulocyte count was done using 6 part fully automated analyser Sysmex (XN-1000 series). Two ml of EDTA blood was collected through a clean venipuncture and Hb electrophoresis was done with mini cap sebia capillary zone electrophoresis. Wherever necessary family study for confirmation was carried out. The results were graphically represented and appropriate statistical software including MS excel, SPSS ver. 20 was used for analysis.

Results

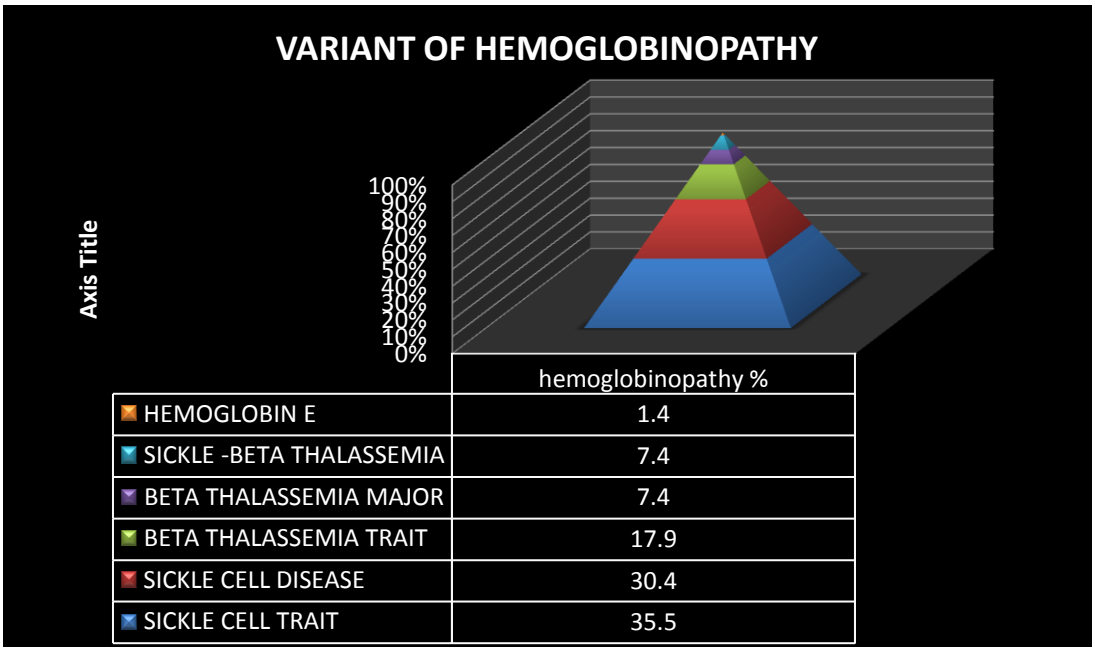
A total of 135 children aged 1 month to 5 years with congenital hemolytic anemia were included in the study.

Graph-1 District-wise demographic Profile of study population. (n= 135)



Graph-1 shows the demographic pattern of the children, maximum cases (46%) belonged to nayagarh district of odisha, followed by khordha (22%).

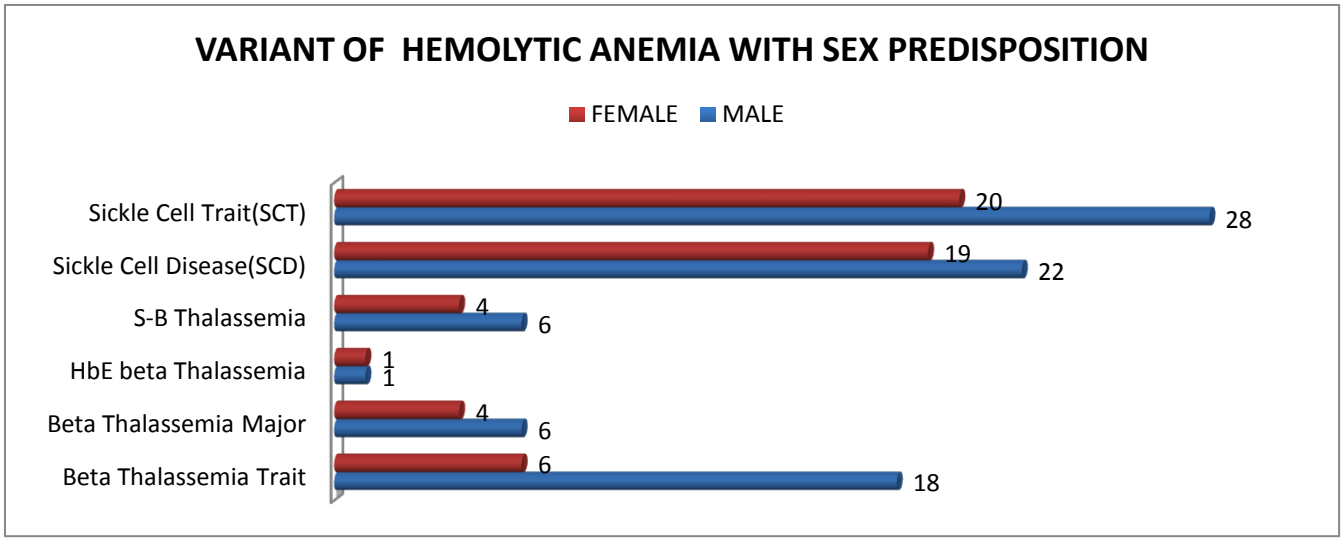
Graph-2 Types of Congenital hemolytic anemia and their prevalence



	hemoglobinopathy %
HEMOGLOBIN E	1.4
SICKLE -BETA THALASSEMIA	7.4
BETA THALASSEMIA MAJOR	7.4
BETA THALASSEMIA TRAIT	17.9
SICKLE CELL DISEASE	30.4
SICKLE CELL TRAIT	35.5

From graph- 2 it was noticed that the most common cause of congenital hemolytic anemia was sickle cell trait (35.5%) followed by sickle cell disease (30.4%), beta thallesemia trait (17.9%), beta thallesemia major (7.4%), S-B thalessemia (7.4%) and hemoglobin E Thalassemia HbE(1.4%)

Graph -3 sex predisposition of Hemolytic anemia



From graph -3 Male are outnumbered female in almost all variety of hemolytic anemia. As the study has less number of population and all geographical strata could not be taken so exact sexual predilection could not be assessed

Table-1 Age distribution according to disease

Age	Beta Thalassemia Major (%)	Beta Thalassemia Trait	HbE Beta Thalassemia	S-B Thalassemia	SCD (%)	SCT (%)
1 to 12 Months	5(50)	6	0	2	6(14.6)	10 (20.8)
1 to 2 Years	2(20)	8	1	1	8(19.5)	13(27)
2 to 5 Years	3(30)	10(41.6)	1	7	27 (65.8)	25(52)
Total	10	24	2	10	41	48

Table 1 shows that majority of patients of sickle cell trait and sickle cell disease belonged to age group of 2 to 5 years with 52% and 65.8% respectively. Beta thalassemia trait was mostly seen in 2-5 years of age (41.6%) whereas beta thalassemia major was observed in 50% of children less than 1 year of age.

Table-2 Hematological Profile of Study Population

Sl No		Beta Thalassemia Trait	Beta Thalassemia Major	HbE	S-B Thalassemia	SCD	SCT
1	Mean Hb(g/dl)	9.58	6.10	8.0	8.50	8.10	11.63
2	Retic Count	0.96	4.50	4.0	3.20	5.44	1.50
3	Mean HbA	88.25	9.50	18.00	14.9	2.51	58.88
4	Mean HbA2	4.42	8.0	4.50	4.4	3.44	3.67
5	Mean HbF	8.38	61.50	11.00	17.6	30.17	6.85
6	Mean Platelet* 1000	259	199	190	63.9	64.2	129.6

As per table-2 The mean hemoglobin was found to be lowest in patients of beta thalessemia major (6.10g/dl). Mean hemoglobin concentration of sickle cell trait was 11.63 so child was having milder form of clinical manifestation and was mainly diagnosed incidentally. Mean reticulocyte count was highest in sickle cell disease. Fetal hemoglobin concentration is more in thalassemia major.

Table-3 Clinical Profile of study population (multiple responses)

Types of Diseases	No of Patients	Percentage(%)
Jaundice	56	41.40
Hemolytic facies	72	53.33
Growth Retardation	67	49.62
Skin Changes	23	17.03
Hepatomegaly	57	42.22
splenomegaly	62	45.92
Edema	7	5.18
Painful crisis	40	29.6

Table 3 shows that maximum percentage of cases had growth retardation (49.62%) followed by splenomegaly and hepatomegaly (45.92%), (42.22%). Hemolytic facies is found approximately 53% of cases in our study.

Discussion

Hemoglobinopathies are prevalent worldwide, but it is more prevalent in certain geographical areas. In the present study, 135 patients of congenital hemolytic anaemia between the age group of 1 month-5 year have been studied clinically as well as by other investigations. The commonest congenital hemolytic anaemia was sickle cell trait 35.5% followed by sickle cell disease and beta thalassemia trait (17.9%) The increased prevalence of HbE Beta Thalassemia in this part of the country was first reported by Chatterjee et al of School of Tropical Medicine, Kolkata in 1966[6]. This higher frequency of HbE Beta Thalassemia can be clarified by the way that these cases having a milder clinical course and consequently presenting at a later age as compared with other patients. Due to its milder clinical course HbE Beta thalassemia contributes least in my study. However as the study population is very small it is difficult to conclude. In this study, the mean hemoglobin was found to be lowest in patients of beta thalassaemia major. Within South Asia, there are about 45 million carriers of beta Thalassemia[7]. Beta thalassaemia among Indian population is seen more commonly in Sindhis, Gujaratis, Bengalis, Punjabis and Muslims[8]. Carrier state for beta thalassaemia in India varies from 1-17% with a mean of 3.2%[9]. Mean hemoglobin concentration of Beta thalassemia major is relatively lesser as compaired to sickle cell counterpart in my study. The incidence of beta thalassemia in my study population is lesser than sickle cell because most form of thalassemia are presented with severe systemic manifestation

and needed PICU care so those patients were excluded from my study. Reticulocyte count increased in most of the congenital hemolytic anemia except beta thalassemia trait (due to ineffective erythropoesis). There were a wide spectrum of clinical manifestations among patients of congenital hemolytic anemia. Symptoms of anemia, pallor, enlargement of frontal, parietal and maxillary bones (hemolytic facies), hepatosplenomegaly associated with jaundice and notched ribs are observed for congenital hemolytic anemia.[10-13] Jaundice (41.4%), hemolytic facies (53.2%) and retarded growth were the common findings in my study. Hepatomegaly (42%) and splenomegaly (45%) were also significant. The major limitation of the study was small sample size and age group within 5 yr . Therefore it is very difficult to correlate the results to general population.

Conclusion

Prevalence of thalassemia varies from state to state, community to community and is a major public health problem .Consanguineous marriage is one of the major contributing factors in which most of the congenital hemolytic anemia are inherited. Children with sickle cell trait have lesser clinical manifestations and may not present to the health care facility early, so community screening among high risk areas should be done.

Genetic counseling, social awareness, education about the disease manifestation, proper prognostication and knowledge about available treatment modalities may be helpful in some extent to reduce the morbidity of the disease. High cost of treatment, repeated blood transfusion, chelation therapy, economic and psychological distress in family, suggest that prevention is better than cure.

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