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Original Research Article

An epidemiological study on the clinico-hematological profile of pediatric patients with congenital hemolytic anemia

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ABSTRACT

Background: Among the inherited disorders of blood, haemoglobinopathy and thalassaemia constitute a major bulk of congenital hemolytic anemia in India.
Methods: The present cross sectional study was conducted on pediatric patients aged 0-15 years admitted in pediatric ward of Department of Pediatrics, MGM Medical College and LSK Hospital, Kishanganj, Bihar, India between December 2015 to November, 2016. The data on socio-demographic profile, relevant clinical history and examination and hematological parameters were assessed.
Results: Out of 211 patients evaluated, most common cause of congenital hemolytic anemia was Hb E Beta thalassemia (39.8%), followed by beta thalassemia (27.9%), beta thalassemia trait (14.2%), Hb E disease (11.3 %) and Hb E trait (6.6%). There was male preponderance (male 63%, female 37 %). The mean hemoglobin was found to be lowest in patients of β thalassemia (5.1 gm/gl) and HbE β thalassemia (5.8 gm/dl). The mean total serum bilirubin was found to be highest among β Thalassemia patients (3.0 mg/dl). Hepatomegaly was the most common clinical finding among the study population (57.8%), followed by splenomegaly (54.9%) and hemolytic facies and jaundice (both 53%).
Conclusions: The incidence of HbE beta thalassemia is relatively high in comparison to other varieties of thalassemias and is a major public health problem in this area of the country.

Keywords: Beta thalassemia, Congenital hemolytic anemia, Hb E beta thalassemia

INTRODUCTION

Haemoglobinopathy and thalassaemia constitute a major bulk of congenital hemolytic anemia in India. They cause a high degree of morbidity, moderate to severe haemolytic anaemia among infants and children and several deaths in India.[1] Congenital hemolytic anemia is anemia due to hemolysis, the abnormal breakdown of red blood cells either intravascular or extravascular. Anemia results If the rate of destruction exceeds the capacity of the marrow to produce red blood cells. Inherited RBC defects of structure and metabolism may result in a chronic hemolytic state, that includes - hemoglobinopathy, like sickle cell anaemia, α thalassemia, β thalassemia, HbE β-thalassemia; RBC enzyme defect, like glucose 6 phosphate dehydrogenase deficiency; RBC membrane disorders like hereditary spherocytosis.[2] Haemoglobinopathies affect 4.5% of the world population.[3]

The prevalence of β- thalassemia trait varies between 3-17% because of consanguinity and caste and area endogamy.[4] Every year, ten thousand children with β-thalassemia major are born in India, which constitutes 10% of the total number in the world.[5] HbE thalassemia is common in north-east parts of India.[6] The only forms

of treatment available for thalassemia patients are regular blood transfusion, iron chelation therapy in an attempt to prevent iron overload and the judicious use of splenectomy in cases complicated by hypersplenism.

The curative treatment like bone marrow transplantation is costly and so prevention is the cost effective strategy, which includes population screening, genetic counseling and prenatal diagnosis.[7]

This study attempts to reveal the clinical and hematological profile of patients with different types of thalassemia admitted in MGM Medical College and LSK Hospital, Kishanganj, Bihar, India.

METHODS

This cross sectional study was carried out in MGM Medical College and LSK Hospital, Kishanganj, Bihar, India over a period of two years from December 2015 to November 2016. Ethical clearance was obtained from the Institutional Ethics Committee and informed consent was taken from the patients.

All the patients admitted in the indoor of the Department of Pediatrics between age group of 0-15 years during the study period were included in the study. Patients with serious systemic illnesses were excluded. All the information regarding their socio-demographic profile, relevant clinical history and clinical examination were collected.

RESULTS

The clinical examination and hematological profile of 211 children with congenital hemolytic anemia have been evaluated in this study. Most common cause of congenital hemolytic anemia was Hb E Beta thalassemia (39.8%), followed by beta thalassemia (27.9%), beta thalassemia trait (14.2%), Hb E disease (11.3 %) and Hb E trait (6.6%) Table 1.

Table 1: Types of congenital hemolytic anemia and their prevalence.

Types	Number	Percentage
Beta thalassemia	59	27.9
Beta thalassemia trait	30	14.2
Hb E beta thalassemia	84	39.8
Hb E disease	24	11.3
Hb E trait	14	6.6

Table 2: Genderwise distribution of congenital hemolytic anemia.

Types	Male (%)	Female (%)
Beta thalassemia	38 (64.4)	21 (35.6)
Beta thalassemia trait	17 (56.6)	13 (43.4)
Hb E beta thalassemia	44 (52.3)	40 (47.7)
Hb E disease	24 (100)	0 (0)
Hb E trait	10 (71.4)	4 (28.6)

Table 3: Hematological profile of congenital hemolytic anemia patients.

Hematological profile		β thalassemia		β thalassemia trait		Hb E β thalassemia		Hb E disease		Hb E trait	
Mean Hb	SD	5.1	0.6	9.2	0.8	5.8	0.6	9.0	0.8	10.1	0.2
Mean TLC	SD	5100	2100	6680	2300	5466	1800	8400	2200	8600	2000
Mean platelet count(×1000)	SD	211	109	243	101	201	96	296	90	290	92
Total serum bilirubin(mg/dl)	SD	3.0	1.6	2.1	1.2	2.8	1.3	2.4	1.1	1.4	0.2

In our study males (63%) were affected more than females (37%).Gender wise distribution in different types of congenital anemia is shown in Table 2. The mean hemoglobin was found to be lowest in patients of β thalassemia (5.1gm/gl) and HbE β thalassemia (5.8 gm/dl). The mean total serum bilirubin was found to be highest among β Thalassemia patients (3.0mg/dl) (Table 3). Hepatomegaly was the most common clinical finding among the study population (57.8%), followed by splenomegaly (54.9%) and hemolytic facies and jaundice (both 53%) (Table 4).

Table 4: Clinical profile of congenital hemolytic anemia patients (multiple responses).

Clinical profile	Number	Percentage
Growth retardation	90	42.6
Jaundice	112	53.0
Hepatomegaly	122	57.8
Splenomegaly	116	54.9
Congestive heart failure	45	21.3
Hmolytic facies	112	53.0
Edema	66	31.2

DISCUSSION

Hemoglobinopathies are prevalent worldwide, but it is more prevalent in some geographical areas. In our study, 211 patients of congenital hemolytic anaemia have been studied clinically an on hematological parameters. The commonest congenital hemolytic anaemia was HbE β-Thalassemia (39.8%) followed by β-Thalassemia (27.9 %). The prevalence of HbE Beta Thalassemia in northeast of the country was reported higher in a previous study.8 This higher incidence of HbE Beta Thalassemia can be explained by the fact that these cases having a milder clinical course and thus presenting at a later age compared to other group of patients of congenital hemolytic anemia, live longer and also get the opportunity to come under medical attention. In our study there was more prevalence of congenital hemolytic anemia in males in respect to females (63 % versus 37%). However it is difficult to conclude as the study sample is small. In this study, the mean hemoglobin was found to be lowest in patients of β thalassemia (5.1gm/gl) and HbE β thalassemia (5.8 gm/dl). The mean total serum bilirubin was found to be highest among β Thalassemia patients (3.0mg/dl). Our results are comparable to previous studies.[9-11] There were a wide spectrum of clinical manifestations among patients of congenital hemolytic anemia. Symptoms of anaemia, pallor, bouts of fever, enlargement of frontal, parietal and maxillary bones (hemolyticfacies), hepatosplenomegaly associated with jaundice and notched ribs are observed for congenital hemolytic anemia.[12-15] Hepatomegaly was the most common clinical finding among the study population (57.8%), followed by splenomegaly (54.9%) and hemolytic facies and jaundice (both 53%). Clinical features of beta thalassemia are usually manifested in younger age group and become more severe with advancing age. HbE beta thalassemia, clinical severity increases with age and complications like those of beta thalassemia eventually develops. Similar results were found in earlier studies.[16,17] The major limitation of the study was small sample size. Therefore it is very difficult to extrapolate the results to general population.

CONCLUSION

Thalassemia is found in almost all the states in India but the prevalence varies from state to state, community to community. The incidence of HbE beta thalassemia is relatively high in comparison to other varieties of thalassemia's and is a major public health problem in this area of the country. Beta thalassemia follows a more severe course and present at younger age compared to other subtypes.

In India, most of the educated people are also not aware of Thalassemia. More efforts are required to bring awareness about thalassemia among the people. High cost of treatment, repeated blood transfusion and chelation therapy and economic burden on family resources, all suggest that prevention is better than cure.

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