Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com/sjams/

OPEN ACCESS

Paediatrics

An Assessment of Clinical Profile and Pattern of Congenital Haemolytic Anemia: A Study in a Tertiary Care Hospital, Rangpur, Bangladesh

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DOI: 10.36347/sjams.2020.v08i02.009 | **Received:** 05.01.2020 | **Accepted:** 12.01.2020 | **Published:** 08.02.2020

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Abstract **Original Research Article**

Introduction: Congenital hemolytic anemia affects a significant proportion of the pediatric population globally. Numerous children are hospitalized every year due to sequelae of this heterogeneous disease. ***Aim of the study*:** The aim of this study was to focus on various aspects of presentation of congenital hemolytic anemia. ***Material & Methods:*** This was a cross sectional study conducted between January 2009 to July 2009 in the Department of Paediatrics, Rangpur Medical College Hospital, Rangpur, Bangladesh. A total number of 40 patients had been included in the study. Base line investigation was done to the new patients admitted in pediatric ward or attending at OPD of Rangpur Medical College Hospital with congenital haemolytic anemia. Case selection was done with complete blood count with peripheral blood film and haemoglobin electrophoresis. ***Results:*** In our study we found the highest number of congenital haemolytic anemic patients were with Hb-E beta thalassemia and it was 65% (n=26). Then case of Hb- E trait was 12% (n=5), case of Hb-E diseases was 10% (n=4) case of beta thalassemia major was 8% (n=3) and case of beta thalassemia minor was 5% (n=2). According to the physical examination of the participants we found, 12.50%, 35% and 7.50% patients had mild, moderate and severe progressive pallor respectively. In total 10% patients were with jaundice whereas 90% were free from that. Again 45%, 22.5% and 10% patients had splenomegaly as < 5cm, 5- 8 cm and >8 cm level. ***Conclusion:*** Nutritional and folic acid supplementation with regular blood transfusion associated with chelating therapy are essential for improving the condition of the diagnosed patients with congenital haemolytic anemia. We would like to recommend for conducting more studies with larger sized sample to get more specific information regarding congenital hemolytic anemia.

Keywords: Congenital Hemolytic Anemia, Pediatric, Thalassemia, Haemoglobin, Electrophoresis.

INTRODUCTION

Congenital hemolytic anemia is a common disease among children, which results mainly from membrane defects, enzyme defects and hemoglobin defects. Again, hemoglobin defects may be due to qualitative (Hemoglobinopathies) and quantitative (Thalassemia syndromes) abnormalities [1]. There is gross geographical and racial variation in the different genes like β or α genes or different hemoglobinopathy genes like Hb-E, Hb-C, Hb-S, etc [2]. The normal haemoglobins are designated by capital letters HbA (or A1) refers to the predominant haemoglobin (more than 95%) found in the normal adults. Hb A2 refers to the normal adult's minor component that constitutes 2.5% to 3.5% of the total haemoglobin. HbF or fetal haemoglobin is the predominant haemoglobin of the fetus or young infants but it presents only in trace amounts (less than 2%) in adult [2]. Haemoglobinopathies such as α and β thalassemia and HbS, HbC and Hb E syndromes are wide spread globally. The Indian subcontinent has a considerably high prevalence of β thalassemia and disorders of Hb E and HbS [3]. Thalassemia is a common disorder with a wide spread geographical distribution. In some region of South –East Asia as many as 40% of the population have one or more thalassemia genes [4]. Bangladesh is a thalassemia endemic zone and around 2000 children are born each year with these diseases in Bangaldesh [5]. The diagnosis of thalassemia is initially suggested by clinical findings variation in ethnic origin and family history and the results of routine haematological profile [6]. Hetarozygotes and homozygotes for Hb-E microytic, minimally anemic and asymptometic. The compound hetarozygosity state of Hb – E beta thalassemia results in a variable and often severe anemia with polypeptide ranging from transfusion

dependence to complete lack of symptoms [7]. Hb-E Beta thalassemia is similar to thalassemia major, anemia is often severe, splenomegaly is almost consistently detected, hemochromatosis is clear, red cell indices and morphology shown microcytosis ,hypochromia, anisocytosis, poikilocytosis, tear drop cell, elliptocytosis target cells and polychromasia are always observed [8]. Congenetial haemolytic anemia has wide spectrum of clinical presentation. It should need considerable clinical studies in all aspects. This study was conducted to focus on various aspects of presentation of the disease.

OBJECTIVES

General Objective

- To evaluate the clinical pattern of congenital haemolytic anemia.

Specific Objectives

- To observe the clinical presentation of congenital haemolytic anemia.
- To assess the clinical presentation of congenital haemolytic anemia.

METHODOLOGY AND MATERIALS

This was a cross sectional study. Conducted in the Department of Peadiatrics, Rangpur Medical College Hospital, Rangpur, Bangladesh. The study was conducted between 1st January 2009 to 31th July 2009. A total number of 40 patients had been included in the study. Base line investigation was done to the new patients admitted in the Paediatric Ward or attending at OPD of Rangpur Medical College Hospital with congenital haemolytic anemia. Case selection was done with complete blood count with Peripheral Blood Film and Haemoglobin Electrophoresis. The inclusion and exclusion criteria were as below:

Inclusion Criteria

- Old and new patients of haemolytic anemia
- Irrespective of previous blood transfusion history.
- Patients of both sexes.
- Age group: 1 – 15 years.

Exclusion Criteria

- Patients suffering from Hemorrhagic disorders or malignancy.
- Diagnosed case of any type of anemia other than congenital haemolytic anemia.
- Not willing to be included in study.

RESULTS

In our study we found the highest number of congenital haemolytic anemic patients were with Hb-E beta thalassemia and it was 65% (n=26). Then case of Hb- E trait was 12% (n=5), case of Hb-E diseases was 10% (n=4) case of beta thalassemia major was 8% (n=3) and case of beta thalassemia minor was 5% (n=2). In all these 40 patients of different types of congenital haemolytic anemia, the age of onset of symptoms varied mostly between 1 to 5 years depending on the nature of the disorders. From the age of onset of symptoms we found, out of 40 cases 15 (37.5%) cases had onset within one year of age of which 8 cases were with Hb-E beta thalassemia, Beta thalassemia major were 1 cases, Beta thalassemia minor were 2 cases, Hb. Besides theses, we found in this study, 2 case of E trait and 2 case Hb- E diseases within one year onset of symptoms also. There were 23 (57.5%) had onset 1 to 5 years of which 16 were with Hb- E beta thalassemia, 3 were with Hb- E trait, 2 were with Hb- E diseases and 2 were Beta thalassemia major. After 5 years of age only 2(5%) cases were found both of them were Hb- E beta thalassemia. In analyzing the features of the patients we found the highest number of patients with progressive pallor and it was 55% (n=22). Then 12.50% had abdominal lump, 10% had abdominal distention, 10% had jaundice, 7.5% had growth failure and 5% had abdominal pain. In our study according to the data of consanguinity we found, in total 15% patients had consanguinity and 85% were free. Among total 6 patients of consanguinity 3 were with Hb-E beta thalassemia, one was with Hb- E trait, one was with Hb-E diseases and another one was with beta thalassemia major. Not a single patient was found with beta thalassemia minor that had consanguinity as well. According to the physical examination of the participants we found, 12.50%, 35% and 7.50% patients had mild, moderate and severe progressive pallor respectively. In total 10% patients were with jaundice whereas 90% were free from that. Again 45%, 22.5% and 10% patients had splenomegaly as < 5cm, 5- 8 cm and >8 cm level. On the other edema was found in 12.5%, ascites was in 7.5%, bony changes were in 40% and growth retardation was found in 45% patients. According to the report of blood transmission of the participants we found 75% patients were regular in blood transmission whereas 25% were irregular. Among all the study people only one patient was found with blood transmitted disease. At the initial stage of the study the hemoglobin concentration was <3 gm/dl, 3- 5 gm/dl, in 7.5%, 5 -8 gm/dl and >8gm/dl in 7.50%, 17.50%, 52.50% and 22.50% patients respectively.

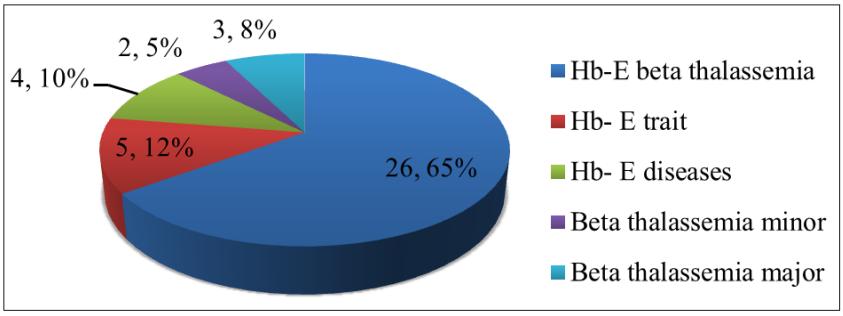

Fig-1: Types of congenital haemolytic anemia (n=40)

Table-1: Age distribution of the participants according to the type of diseases (n=40)

Disease Type	<1 Year	1-5 Years	>5 Years	Total	%
Hb-E beta thalassemia	8	16	2	26	65.00
Hb- E trait	2	3	0	5	12.50
Hb- E diseases	2	2	0	4	10.00
Beta thalassemia minor	2	0	0	2	5.00
Beta thalassemia major	1	2	0	3	7.50
Total	**15**	**23**	**2**	**40**	**100**

Table-2: Feature distribution among the participants (n=40)

Presenting complaints	**n**	**%**
Pallor	22	55.00
Abdominal distention	4	10.00
Abdominal Lump	5	12.50
Jaundice	4	10.00
Growth Failure	3	7.50
Abdominal Pain	2	5.00
Total	40	100

Table-3: Status of consanguinity among the participants (n=40)

Disease Type	**Total**	**Consanguinity**	
		Yes	**No**
Hb-E beta thalassemia	26	3	23
Hb- E trait	5	1	4
Hb- E diseases	4	1	3
Beta thalassemia minor	2	0	2
Beta thalassemia major	3	1	2
Total	**40**	6 (15%)	34 (85%)

Table-4: Physical examination of the study subjects. (n=40)

Physical signs		**N**	**%**
Pallor	Mild	5	12.5
	Moderate	14	35
	Severe	3	7.5
Jaundice	Absent	36	90
	Present	4	10
Hepatomegaly	Not palpable.	9	22.5
	< 5cm	18	45
	5- 8 cm	9	22.5
	>8 cm	4	10
Splenomegaly	Not palpable	4	10
	< 5cm	12	30
	5- 8 cm	17	42.5
	>8 cm	7	17.5
Edema	Absent	35	87.5
	Present	5	12.5
Ascites	Absent	37	92.5
	Present	3	7.5
Bony changes	Absent	24	60
	Present	16	40
Growth retardation	Absent	22	55
	Present	18	45

Table-5: Hemoglobin concentration of the study subjects at initial presentation (n=40)

Hemoglobin range	N	%
<3 gm/dl	3	7.50
3- 5 gm/dl	7	17.50
5 -8 gm/dl	21	52.50
>8gm/dl	9	22.50

DISCUSSION

In this study total number of 40 cases of congenital haemolytic anemia in children less than 15 years of age were studied for a period of 6 months in the Department of Paediatrics in Rangpur medical college hospital to evaluate the pattern and clinical profile of these children with existing facilities in locality. In different types of thalassemia syndrome Hb E- beta thalassemia in the most prevalent disorder in our country. Here out of 40 cases 26 cases (65%) were Hb E beta thalassemia. 5 cases (12%) were Hb-trait and 4 cases (10%) were Hb–E disease. Hb-E is more prevalent in this study group. This is because Hb-E is common abnormal haemoglibin in this region of the world [14]. It is more common in South-East Asia, India, Myanmar and Srilanka. About 13% population of Thailand [15], Cambodia and Laos being affected. About 50% of population in Saudi Arabia carries the gene of Hb-E [9]. Therefore the results in this study is consistent with the findings of other study. In this study 2 cases (5%) of thalassemia major and 3 cases (8%) of thalassemia minor were found. This is of course more common in Greece, Italy and other Mediterranean countries and also Negroes of Americal stock [10]. But it is not absent in this part of the world and infact thalassemia major is prevalent in South-East Asia [11]. Findings of this study suggests thalassemia major in not uncommon in this part of the world and revealed the existence of beta thalassemia major in Bangladesh as in previous study [16]. In this study no cases of α thalassemia was found. Alpha thalassemia as well as with compound heterozygote is common in Southeast Asian countries [12]. Saudi Arabia is one of the countries with the highest incidence of alpha-thalassemia [2]. But both α and β thalassemia, Hb-E and Hb constant spring are common in Thailand [13]. There was no case of sickle cell anemia found in this study, though it had been reported in this country [17]. Other types of congenital haemolytic anemia could not be included in this study due to lack of investigation facilities. In all these 40 patients of different types of congenital haemolytic anemia, the age of onset of symptoms varied mostly between 1 to 5 years depending on the nature of the disorders. In all these 40 patients of different types of congenital haemolytic anemia, the age of onset of symptoms varied mostly between 1 to 5 years depending on the nature of the disorders. From the age of onset of symptoms we found, out of 40 cases 15 (37.5%) cases had onset within one year of age of which 8 cases were with Hb-E beta thalassemia, Beta thalassemia major were 1 cases, Beta thalassemia minor were 2 cases, Hb. Besides theses, we found in this study, 2 case of E trait and 2 case Hb- E diseases within one year onset of symptoms also. There were 23 (57.5%) had onset 1 to 5 years of which 16 were with Hb- E beta thalassemia, 3 were with Hb- E trait, 2 were with Hb- E diseases and 2 were Beta thalassemia major. After 5 years of age only 2(5%) cases were found both of them were Hb- E beta thalassemia. According to the physical examination of the participants we found, 12.50%, 35% and 7.50% patients had mild, moderate and severe progressive pallor respectively. In total 10% patients were with jaundice whereas 90% were free from that. Again 45%, 22.5% and 10% patients had splenomegaly as < 5cm, 5-8 cm and >8 cm level. On the other edema was found in 12.5%, ascites was in 7.5%, bony changes were in 40% and growth retardation was found in 45% patients. Signs vary according to the severity and duration. This indicates growth retardation is invariably present in thalassemia major, and not invariably present in Hb-E beta thalassemia patients, though chronic haemolytic anemia is one of the causes of growth retardation. This is probably because many patients in this study followed advices of regular blood transfusion but properly not maintain and other measures. Among all the study people only one patient was found with blood transmitted disease. At the initial stage of the study the hemoglobin concentration was <3 gm/dl, 3- 5 gm/dl, in 7.5%, 5 -8 gm/dl and >8gm/dl in 7.50%, 17.50%, 52.50% and 22.50% patients respectively. Radiological findings revealed hair on end appearance of the skull bone in several cases of thalassemia major and Hb–E beta thalassemia cases who had been suffering from early period of life. This represents that gross bony changes occurs in the long-standing cases. Serum iron and ferritin level could not be measured due to lack of facilities locally and Result of treatment and relation with blood group had not been included in this study.

LIMITATIONS OF THE STUDY

This was a prospective type of study in a single community with comparatively small number of sample size. So, the study result may not reflect the exact scenarios of the whole country.

CONCLUSION AND RECOMMENDATIONS

From the current study, we can conclude that Hb –E β thalassemia is the most prevalent disorder in our country and they are presented as early as before the one year of age. Progressive pallor, jaundice, abdominal distention, growth retardation, bony changes are and hepatosplenomegaly are common presenting features. Nutritional and folic acid supplementation with regular blood transfusion associated with chelating therapy are essential for improving the condition of the disease process. Proper screening method should be applied to reduce it as well as carriers with premarital genetic counseling. Wide spectrum of study needed, prenatal diagnosis should be arranged to prevent mortality and morbidity of the disorder.

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