

Original Research Paper

Paediatrics

CLINICO-HAEMATOLOGICAL PROFILE OF HEMOLYTIC ANEMIA IN CHILDREN

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ABSTRACT

Background: The purpose of study is to evaluate clinical profile of children with hemolytic anemia with necessary investigation along with growth parameters of these children with reference to anemia and parental awareness to recognize the degree of pallor in their affected children.

Methods: The study was carried out in patients of Krishna hospital and Medical research centre, Karad in period of January 2018 to August 2018. A total of 36 cases were included in the study. Children aged between 4 months to 12 years diagnosed to have hemolytic anemia, those who had given consent, those who came to our hospital for blood transfusion were included in study.

Results: Among the study, sickle cell disease , sickle beta thalassemia and sickle beta thalassemia trait are present in majority of the patients (61.09%). Beta thalassemia major along with beta thalassemia intermedia is present in 38.88% of the cases. Conclusion: Sickle beta thalassemia and sickle cell disease forms the major types of hemolytic anemia .Parental awareness of $presence\ of\ paleness\ and\ anemia\ is\ poor.\ No\ relation\ to\ religion\ ,\ social\ group\ and\ consanguinity\ .$

KEYWORDS: Thalassemia, Hb electrophoresis, serum ferritin, pallor.

INTRODUCTION:

Hemolysis is defined as the premature destruction of red blood cells. Anemia results when the rate of destruction exceeds the capacity of the marrow to produce RBCs.

Essential feature of hemolytic anemia is reduction of normal red cell survival (110-120 days). Premature destruction of red blood cells may result from:

- Corpuscular abnormalities: Membrane defects, enzyme defects and haemoglobinopathies.
- Extra- corpuscular abnormalities: Immune-mediated, auto-immune, infections and drugs.

The World Health organizations (WHO) has suggested that about 5% of the world population are carriers of different inherited disorders of haemoglobin.

For optical management of Hemolytic anemia, a multidisciplinary approach is essential involving Pediatrician, Haematologist, Genetist, Transfusion specialist, endocrinologist, Child psychiatrist, and others. Poor infrastructure, lack of facilities, lack of knowledge have resulted in failure of community control of birth of these totally preventable dreadful genetic disorders. Prenatal diagnosis is one of the important aspects of prevention programme. There is a dilemma in counseling couples for prenatal diagnosis for sickle cell syndromes because if the course of the disease is mild, it is unnecessary to go for prenatal diagnosis. Awareness about the disorder should be generated for successful prevention programme.

Ethical Statement: The Study made the standards outlining the declaration of Helsinki and Good Epidemiological practices. This study did not change or modify the laboratory of clinical practices of each centre and differences of practices were kept as they are. The data collection was anonymous and identifiable patient information was not submitted.

Individual researchers were responsible for complying with local ethical standards and hospital registration of study.

Methods of collection of data:

Inclusion criteria:

Children aged between 4 months to 12 years diagnosed to have haemolytic anemia.

- Who had given consent to take part
- Who came to our hospital for blood transfusions.

Exclusion criteria:

- Whose parents did not agree to take part
- Below 4 months and above 12 years of age.
- With other co-morbid conditions like cardiomyopathies, HBsAg or HCV positive or with severe systemic illness.

Sample Size: A total of 36 cases were included in the study.

Method of Examination:

- Explaining objectives of the study
- An informed consent after proper counseling
- Detailed history to be taken.
- Transfusion history to be taken
- A detailed comprehensive physical examination.
- Blood samples and imaging studies done

Investigations done:

- Complete blood count, red cell indices, liver function tests, peripheral smear, and reticulocyte count.
- Hb electrophoresis
- Serum ferritin estimation
- HIV I and II, HBV, HCV.

OBSERVATION AND RESULTS:

The study was carried out in patients of Krishna hospital and Medical research centre, Karad in period of January 2018 to august 2018. A total of 36 cases were included in the study.

Table 1: Distribution of children according to age

Age in years	No. of children	Percentage(%)
0.33-0.92	11	30.55
1.00-3.00	11	30.55
4-8	9	25.00
10-12	5	13.88
Total	36	100

Chart 2: Distribution of children according to sex Distribution of children according



Table 2: Distribution of children according to religion

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Religion	No of children	Percentage (%)		
Hindu	29	80.55		
Christians	7	19.45		
Total	36	100		

Chart 2: Distribution of children by social group

Distribution of children by social group

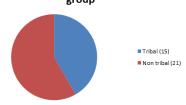


Table 3: Distribution of Consanguinity

Consanguinity	Present	Absent
Number	15	21
Percentage	41.67%	58.33%

Table 4: Distribution of children by Haemoglobin Group

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HB group	< 5 gm/dl	5-10 gm/dl	total
Number	21	15	36
Percentage	58.33%	41.67%	100%

Chart 3: Recognition of paleness of children by Parents
Recognition of paleness of children
by Parents



Table 5; clinical presentation of cases

Clinical features	number	Percentage(%)
pallor	36	100
icterus	17	47.22
cardiomegaly	5	13.89
Cvs – flow murmer	20	55.56
respiratory	12	33.33
crepitations	11	30.56
Decreased breath sounds	1	2.78
hepatomegaly	32	88.89
splenomegaly	34	94.44
Splenic tenderness	2	5.56
Hemolytic facies	8	22.22

Chart 4: Types of Hemolytic Anemias

|| sickle ce || disease(7) || sickle beta thalassemia(12) || sickle beta thalassemia trait (2) || sickle ce || trait(1) || beta thalassemia major(13) || beta thalassemia intermedia(1)

Types of Hemolytic Anemias

Among the study, sickle cell disease, sickle beta thalassemia and sickle beta thalassemia trait are present in majority of the patients (61.09%). Beta thalassemia major along with beta thalassemia intermedia is present in 38.88% of the cases.

DISCUSSION AND SUMMARY:

The study titled "Clinico-haematological profile of Haemolytic anemia in Children" was undertaken with the following objectives.

- To find the pattern of haemolytic anemia in the study
- To correlated clinical and haematological findings.

- A total of 36 children were included in the study with age range from 4 months to 12 years. A combined evaluation of clinical presentation, physical findings and routine haematological investigations and haemoglobin electrophoresis were carried out.
- Age distribution 61% of cases are below 2 years of age. Out of which 30.55% are below one year of age.
- Sex distribution majority of the children were males about 62% of the total cases.
- Religion distribution largest group of these cases were among Hindus.
- Consanguinity interestingly 58% cases had no obvious consanguinity though haemolytic anemia is a genetic disorder where consanguinity is seen often.
- Significant family history was present in 11.11% of cases.
- Growth patterns in children- almost all cases were below normal that is less than 50th percentile for weight for age and nearly 2/3rd of the cases were below 3rd percentile for weight for age there was significant growth retardation in the children.
- Majority of the children 58.33% had severe anemia having haemoglobin less than 5gm/dl
- Clinical presentation- pallor is present in all the cases.
 Splenomegaly is seen in 95% of cases, hepatomegaly in 88.89% and icterus in 47.22% of cases.
- Haematological characteristics- majority of the cases showed microcytic hypochromic blood picture with anisocytosis, target cells and sickle cells. Sickling test was positive in all children with sickle cells disorders. Majority of the cases had reticulocyte count more than 2% so this value can be taken as a cut off for the suspicion of haemolytic anemia there by helping in the early diagnosis and management of these case.
- Haemolytic anemias observed in this study were sickle beta thalassemia (33.33%) sickle cell Disease (19.44%), sickle cells that (2.77%) sickle beta thalassemia trait (5.55%) beta thalassemia major (36.11%) and beta thalassemia intermedia (2.77%)
- Sickle cell anemia and sickle beta thalassemia contributing 53% of the cases.
- One case of heterozygous haemoglobin SD (HbSD) has presented with pallor and weakness and confirmed by haemoglobin electrophoresis.
- Beta thalassemia major contributes to 36.11% of the cases.
 And beta thalassemia intermedia are present in 2.77% of the cases.
- Among children with Beta thalassemia major, 6 children are on oral chelation therapy with regular blood transfusions and no transfusion reactions were noted.
- No Transfusion related reactions or complications were found in the children during blood transfusions.

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