Original Research Paper

Volume - 10 | Issue - 11 | November - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Paediatrics

PATTERN AND CLINICAL PROFILE OF PATIENTS WITH B- THALASSEMIA IN REPEATED BLOOD TRANSFUSION

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ABSTRACT

Background: Chronic transfusions inevitably lead to iron overload as humans cannot actively remove excess iron. The cumulative effects of iron overload lead to significant morbidity and mortality, if untreated. The combination of transfusion and chelation therapy has dramatically extended the life expectancy of thalassemia patients, but with complications like hypocalcaemia. Hence, present study was undertaken to determine pattern and clinical profile of patients with β- thalassemia who are receiving repeated blood transfusion

Methods: Hospital based study conducted at S. Nijalingappa Medical College and Hanagal Shri Kumareshwar hospital, Bagalkot. The study period was one and half year from 2015 to 2016. 53 beta thalassemia major cases fulfilling inclusion criteria were investigated after an informed consent, for serum calcium, serum phosphorous, serum ALP and paratharmone levels.

Result: Among 53 transfusion dependent children studied, the mean age is 5.249 years. The study consisted of 32 (60.4%) males and 21 (39.6%) females. Maximum number of cases i.e. 29 (54.7%) were diagnosed at the age of 4-6 months. 50 (94.3%) were on iron chelation therapy. The mean serum calcium is 8.28 $\pm$ 0.89 mg/dl. The mean serum phosphate is 6.40 $\pm$ 0.80mg/dl, mean PTH is 14.96 $\pm$ 15.49ng/L. The mean value of serum phosphate level is 14.96 $\pm$ 15.49 ng/L. The mean ALP is 166.789 U/L.

Conclusion: To get better results, regular testing is needed to detect the complications of the early stages with proper treatment of the factors and complications. Therefore, should be monitored to avoid complication related to hypocalcemia.

KEYWORDS : Beta- thalassemia major, Blood transfusion, Chelation therapy, Children

INTRODUCTION:

Thalassemia is usually treated via blood transfusion to provide the patients with healthy red blood cells containing normal hemoglobin [1].Although blood transfusions are important for patients with anemia, chronic transfusions inevitably lead to iron overload as humans cannot actively remove excess iron. The cumulative effects of iron overload lead to significant morbidity and mortality, if untreated. A unit of red blood cells transfused contains approximately 250 mg of iron, while the body cannot excrete more than 1 mg of iron per day [2]. However, repeated blood transfusions can lead to iron overload, whereby excess iron accumulates in the body and is deposited in body organs such as the heart, liver and endocrine glands causing organ damage [1].

Iron overload is the main problem encountered in the management of thalassemia. Attributed to excessive iron overload and suboptimal chelation, endocrine dysfunction is a common complication of beta-thalassemia. Despite a significant increase in the lifespan of these patients, many endocrine abnormalities such as hypogonadism, diabetes mellitus, hypothyroidism and hypoparathyroidism (HPT) develop due to an iron overload [3].

Over the past three decades, regular blood transfusions and iron supplementation have improved the quality of life and transformed the major Thalassemia from a rapidly fatal disease in early childhood to chronic diseases compatible with prolonged life. Available treatments for patients with thalassemia are routine blood transfusions, iron therapy for the purpose of preventing iron overload, and the judicious use of splenectomy in cases complicated by hypersplenism, and hematopoietic stem cell transplantation. Patients with beta-thalassemia major are prone for hypoparathyroidism; an irreversible and preventable disorder caused by iron overload [4]. This condition is asymptomatic in most β thalassemia patients, with hypocalcemia usually only detected during routine laboratory examinations [5]. Therefore, the present study was undertaken to determine pattern and clinical profile of patients with β- thalassemia who are receiving repeated blood transfusion.

MATERIAL AND METHODS:

This prospective study was conducted in the Department of Paediatrics, SNMC & HSK Hospital and Research centre Bagalkot from 1.12.2015 to 30.11.2016 after taking approval from the review board of Institutional Ethical Clearance committee. For every patient, detailed assessment of history, clinical examination, investigations, treatment, and follow-up were performed.

INCLUSION CRITERIA :

Patients diagnosed with β- thalassemia major by electrophoresis, Receiving repeated blood transfusion, Serum ferritin levels more than 1000ng/ml.

EXCLUSION CRITERIA :

Poor compliance for blood transfusion, On any drug which affect the calcium level, Having renal disease, Malabsorption syndrome, Rickets.

Data collection procedure:

Patients diagnosed with β-thalassemia major, who are receiving regular blood transfusion, were selected for the study. They were examined for the signs like chvostek sign and trousseau sign. Their anthropometry was done. Informed written consent was taken from guardians of the patients.

For the collection of data, pre-prepared proforma was used which included personal information, data on the number of transfusions and the inclusion of hemoglobin and serum ferritin.

The current study collected data using a pre-designed form that served the purpose of the study. In-depth physical examination, including anthropology, general examination and systematic examination was performed and recorded in proforma documents. Serum ferritin levels were measured in all patients with thalassemia. Iron chelating agents are recommended for all patients with serum ferritin levels in excess of 1000 ng / ml.

4ml venous blood was taken, 2 ml in plain bulb and 2 ml in EDTA bulb. EDTA bulb is sent for hemogram. Serum calcium was estimated using o-Cresolphthalein (oCPC method). Ammonium molybdate method was employed to estimate serum phosphate levels. And chemiluminescent immunoassay (CLIA) method was used to estimate the parathormone levels.

STATISTICAL ANALYSIS:

Descriptive statistics such as mean, SD and percentage was used to present the data. Data were entered in MS Excel and analyzed in SPSS Version 22.

RESULT:

Table 1 : Basic characteristics

Characteristics	Number	Percentage
Age		

<1	5	9.4
1-5	28	52.8
6-10	14	26.4
11-14	6	11.3
Gender		
Male	32	60.4
Female	21	39.6
Age at diagnose (in months)		
≤ 3	9	17.0
4-6	29	54.7
7-9	10	18.9
10-12	5	9.4

Among 53 transfusion dependent children studied, majority i.e. 28 (52.8%) of the children belonged to the age group of 1-5 years. The maximum age studied was 14 years of age. Among the cases studied, the mean age was found out to be 5.249 years. The study consisted of 53 thalassemia patients among which, 32 (60.4%) were males and 21 (39.6%) females. Maximum number of cases i.e. 29 (54.7%) were diagnosed at the age of 4-6 months.

Table 2 : Transfusion pattern

	Number	Percentage
Number of transfusions		
≤ 25	12	22.6
26-50	13	24.5
51-75	8	15.1
76-100	8	15.1
>100	12	22.6
Frequency of transfusion (in days)		
15	8	15.1
30	45	84.9

These transfusion dependent children studied had mean number of transfusion of 68.11. Among the thalassemia patients studied, majority of them i.e. 45 (84.9%) needed packed red blood cells transfusion every 30 days. Whereas, very few i.e. 8 (15.1%) children required transfusion every 15 days.

Table 3: Clinical profile

	Number	Percentage
On iron chelation therapy	50	94.3
Stunting	27	50.9
Chvostek sign	25	47.2
Trousseau sign	9	17.0
Hepatomegaly	51	96.2

Among 53 transfusion dependant patients, 50 (94.3%) were on iron chelation therapy. Stunting was observed in 27 (50.9%) of these children. 25 children had chvostek sign positive, whereas the rest 28 did not have chvostek sign. 17% children had shown Trousseau sign. Majority of the children 96% showed hepatomegaly.

Table 4: Investigation parameters

Parameters	Mean	SD
Serum ferritin ()	1971.15	725.60
Serum calcium (mg/dl)	8.28	0.89
Serum phosphate (mg/dl)	6.40	0.80
Serum PTH (ng/L)	14.96	15.49
Serum alkaline phosphatase (U/L)	166.78	25.24

Children with serum ferritin more than 1000 were studied, among which the maximum serum ferritin level observed was 3657 and mean level of 1971.15 with standard deviation of 725.60. The serum calcium levels of the cases studied ranged from 6.30 mg/dl to 11.50 mg/dl. The mean value of serum calcium level was found to be 8.28 ± 0.89 mg/dl.

The serum phosphate levels of the cases studied ranged from 2.40mg/dl to 6.40 mg/dl. The mean value of serum phosphate level was found to be 6.40 ± 0.80 mg/dl. The serum PTH levels of the cases studied ranged 5.40 to 105 ng/L. The mean value of serum phosphate level was found to be 14.96 ± 15.49 ng/L. The serum ALP ranged from 106 U/L to 289 U/L with the mean value of 166.789 U/L.

DISCUSSION:

Age distribution

The age group of patients in the present study was between 1-5 years, with the mean age being 5.249± 3.4562 years, comparable with other study with mean age 6.35 ± 2.27 years [6]

Sex distribution

The reason for current male preponderance in Indian studies could be attributed to the Indian society where the female children are neglected. There is no reason for the male preponderance in thalassemia major. Findings of present study was comparable with study done by George (60%) and Choudhary VP (54.9%) [7,8].

Age of diagnosis

In this study the majority of the cases i.e 54.7% was diagnosed at the age of 4-6months. β thalassemia major manifests very early childhood with pallor being predominant with in the first year of life.

Frequency of transfusion

Lokeshwar MR, et al states that transfusion to be given at the interval of 2-4 weeks [9]. In the present study, it was observed that majority of the thalassemia patients i.e. 84.9% needed transfusion every 30 days (4 weeks) whereas, only 15.1% of them needed transfusion for every 15 days (2 weeks).

Investigation parameters:

Serum calcium

Aleem AA, et al in their study observed ionized calcium levels in thalassemia patients and found the levels low, ranging from 1.58mmol/L – 2.04mmol/L and a mean value of 1.88mmol/L [10].

Hagag AA, et al also studied about the ionized calcium levels in β-thalassemia patients and concluded that the ionized calcium levels were as low as 0.60-1.23

mmol/L with a mean value of 0.96±0.13 [1].

In the present study, patients with β- thalassemia have low serum calcium levels ranging from 6.30-11.50 mg/dl and a mean value of 8.28±0.89 mg/dl.

Findings of the present study was comparable with studies done by Goyal M with 8.42±0.32, Modi AS with 8.37±0.20 [11,12].

Serum phosphate

In the present study, serum phosphate levels were observed to be ranging from 2.40-6.40mg/dl with the mean value of 4.19±0.80 which was found to be increased. This suggests that serum phosphate levels were found to at the higher level of the normal range. Findings of the present study was comparable with the study done by Modi AS with 4.87 ± 0.37, Angelopoulos NG with 4.87 ± 1.39 [12,13].

Serum PTH

In the present study parathormone levels (14.96 ± 15.49) were found to be low which was also seen in studies done by Goyal M,et al (32.2 ± 0.96) and Angelous NG,et al. (10.48±4.46) [11,13], whereas Basha KP, et al study did not find any clear relation between serum ferritin and serum PTH [14].

Alkaline phosphatase

Goyal M, et al in their case control study studied the alkaline phosphatase levels in the thalassemia major patients who received blood transfusion and compared with the normal children. They found the levels of ALP in patients to be 7.67± 0.85 (KA U/L) [11]. In the present study, the mean value of ALP is found to be increased.

CONCLUSIONS

Thalassemia major patients significantly showed spleenomegaly and hepatomegaly. To get good results, regular testing is needed to detect the complications of the early stages with proper treatment of the factors and complications. Therefore, should be monitored to avoid complication related to hypocalcemia.

REFERENCE:

1. Hagag AA, Elfrargy MS, Gazar RA, Abd El-Lateef AE. Therapeutic value of combined therapy with deferasirox and silymarin on iron overload in children with beta thalassemia. Mediterranean journal of hematology and infectious diseases. 2013;5(1).
2. Ikram N, Hassan K, Younas M, Amanat S. Ferritin levels in patients of Beta Thalassemia major. Int Juornal Pathol. 2004;2(2):71–4.
3. Jensen CE, Tuck SM, Agnew JE et al. High prevalence of low bone mass in thalassaemia major. Br J Haematol. 1998 Dec; 103(4): 911-5.
4. Hamidieh AA, Moradbeag B, Pasha F, Jalili M, Hadjibabaie M, Keshavarznia M. Hypoparathroidism in patients with beta thalassemia major. IJHOSCR. 2009; 3:17-20.
5. Wieliczko M, Dylewska M. Hypocalcemia. Wiadomosci Lekarskie (Warsaw, Poland: 1960). 2013 Jan 1;66(4):303-6.
6. Anju R, Jain S. Study of Serum Calcium and Serum Phosphorus Levels in Patients of Thalassemia Receiving Repeated Blood Transfusion.2017;6(1):2015–7
7. George A,Bhaduri A, Sen S and Choudhary VP. Physical growth parameters in thalassemic children. Indian J Pediatrics 1997;64:861-71.
8. Choudhry VP, Minor T. Thalassemia Minor and Major : Current Management. The Indian Journal of Pediatrics; 2017;(1):34-6.

9. Lokeshwar M R, Manglani M, Shaw N, Kanakia S:Modern trends in the management of thalassemia, Indian Journal of Pediatrics 1997;5:287-95.
10. Aleem A, Al-Momen A-K, Al-Harakati MS, Hassan A, Al-Fawaz I. Hypocalcemia due to hypoparathyroidism in (beta)-thalassemia major patients. Ann Saudi Med. 2000;20(5):364–6
11. Goyal M, Abrol P, Lal H. Parathyroid and calcium status in patients with thalassemia. Indian J Clin Biochem. 2010;25(4):385–7.
12. Modi AS, Poornima RT, Jayaprakash Murthy D. Serum calcium and phosphate levels in patients with β-thalassemia major. IJPBS 2012;2(4):156-60.
13. Angelopoulos NG, Goula A, Rombopoulos G, Kaltzidou V, Katounda E, Kaltsas D, et al. Hypoparathyroidism in transfusion-dependent patients with b - thalassemia. 2006;138–45.
14. Basha K parveen, Shetty B, U.V.Shenoy. Hypoparathyroidism in beta- thalassemia major. Acta Haematol. 1992;88(2):105–8.