



Study Of Frequency Of Blood Transfusion And Hematological Profile of the Known Cases Of Beta -Thalassemia Major Patients Attending At Tertiary Care Hospital

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Abstract:

Introduction:

Beta thalassemia syndromes are the group of disorder which is hereditary (Autosomal Recessive) with characterized by reduced or absent beta globin chain synthesis resulting in the reduced hemoglobin in the Red blood cell (RBC), Decreased RBC production and anemia.

Material and Methods:

We studied the 50 patients attending the blood bank for transfusion from the pediatric department of C.U.Shah medical College and hospital, Surendranagar from October 2022 to September 2023.

Aims and objectives:

To determine the age of patients, at the age when they are diagnosed, frequency of transfusion, total number of transfusions till the study is done and correlation with The serum Ferritin level, hemoglobin level and MCHC is also studied from records.

Result:

Among 50 patients, mean age is 9.48 years, with the mean duration 8.25 years till study done having mean hemoglobin up to 7.04 gm/dl, mean level of MCHC is 36.48 g/dl, due to once or twice a month transfusion to maintain the serum ferritin level, majority are on the chelation therapy (96%), mean Serum ferritin level is correlated with the number of transfusion in <150 is 2948±1745.69 and in >150, 3100±1365.42 microgram/dl. Male to female ratio is 1.38 in the study done.

Conclusion:

It is mandatory to monitor the hemoglobin level, serum ferritin with the transfused units to give them a better and prolonging life expectancy.

Key words: Beta thalassemia, Transfusion, Chelation

I Introduction :

Beta thalassemia syndromes are the group of disorder, which is hereditary (Autosomal recessive) with characterized by reduced or absent beta globin chain synthesis, resulting in the reduced hemoglobin in Red blood cells (RBC), Decreased RBC production and anemia. Hemoglobin is the tetramer of two alpha and two beta chains. When there is disturbance in the level of either alpha or beta, causing a syndrome called thalassemia. In beta thalassemia beta chain is either decreased or not synthesized basis on that the three conditions of increasing severity are recognized, i.e., beta thalassemia carrier state, thalassemia intermedia and thalassemia major.^[1] Data are still lacking for many regions of the world ,recent data indicates that about 7%of the world's population is a carrier of a hemoglobin disorder and that 3,00,000 to 5,00,000 children are born each year with the severe homozygous state of the world wide^[1] The carrier rate for thalassemia genes varies from 3 to 15% in northern India and 1 to 3 %in southern India.^[2] Every Approximately 1,00,000 are born with thalassemia in India.^[3] The screening of the thalassemia is by Complete blood count, the confirmatory test is the hemoglobin electrophoresis. Beta thalassemia major is transfusion dependent, which requires the packed Red cell regularly. The effective and good transfusion regimen is required for (1)Good growth and development (2)Good energy level and (3) Sufficient suppression of intra and extramedullary hematopoiesis^[4,5] Monitoring of the Hemoglobin level is mandatory for the management aspect. To reduce the iron overload after such blood transfusion units serum ferritin level should be done after every 3 months and chelation agent is required for the maintaining the serum ferritin level 500-1000 microgram/liter.^[6,7]

II. MATERIALS AND METHODS:

In the present study, we studied the total 50 patients who are attending the blood bank for the blood transfusion from the pediatric department at tertiary care hospital, surendranagar from October 2022 to September 2023. The objective of the study is to assess the blood transfusion profile and their outcome. We have been interviewed the patients for their clinical history regarding the Age, at what age they diagnosed as a Beta thalassemia major , duration of disease,frequency of transfusion,total number of transfusions up to the study done along with that for to study purpose we taken up the clinical and laboratory investigation records of hemoglobin level, MCHC,serum ferritin level. The obtained data is analyzed and expressed in terms of mean, median, mode, ratio, percentage, mean \pm standard deviation (SD).

III. RESULTS AND DISCUSSION

Results of Descriptive Statics of Study Variables

Table 3.1: Comparison of the variables from different subgroups to the previous study in percentage

Variable	Subgroups	Total numbers	Percentage	Comparison with other studies (Pattanashetti et al.) ^[8]
Age at diagnosis(Months)				
	6 or less	10	20%	8.57
	7-12	26	52%	45.71
	13-18	13	26%	42.86
	>18	01	02%	2.00
Total		50	100.00	100.00
Duration of disease (Years)				
	1-5	10	20	20.00
	6-10	19	38	20.00

	11-15	17	34	40.00
	16-20	04	08	17.14
	>21	00	00	00.00
Total		50	100.00	100.00
Frequency of transfusion(Per month)				
	1	34	68%	91.43
	2	16	32%	8.57
Total		50	100.00	100.00
Total number of transfusion				
	<50 51-100	14 03	28% 06%	5.71
	101-150	17	34	51.43
	151-200	08	16	25.71
	201-250	01	02	14.28
	>250	07	14	2.86
Total		50	100.00	100.00
Chelation Therapy				
	Yes	48	96.00	71.43
	No	02	04.00	28.57
Total		50	100.00	100.00

TABLE 3.2: Age of the patients in years

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Age (Years)	9.48	4.55	10	1-20

The mean age of the patients is 9.48 ± 4.55 years and median is 10 years , in which the oldest one is of 1year and maximum is of 20 years. Compared with the previous study done was pattanashetti et.al

^[8]In which the mean age is 13.46 ± 3.67 years .

TABLE 3.3: Age at the diagnosis done as a Beta thalassemia major

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Age at diagnosis(Months)	19.22	21.05	12.00	1-96

The mean age at which they diagnose Beta thalassemia major is 19.22 ± 21.05 months with median is 12 months, in which the minimum age at diagnosis is 1month to maximum is 96 months. We compare the study done with pattanashetti et al. is 8.94 ± 6.15 months.

TABLE 3.4: Duration of disease from the diagnosis till the study done

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Duration of disease (Years)	8.25	4.57	8.5	0.5-16.5

The mean of the duration of disease is 8.25 ± 4.57 years ,median is 8.5years and minimum is 0.5 to maximum is 16.5 years . Compared to other previous studies, pattanashetti et al. Mean is 12.29 ± 3.66 years .

TABLE 3.5 : Frequency of transfusion required in one month

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Frequency of transfusion (Per month)	1.32	0.46	1	1-2

The mean frequency of transfusion is 1.32 ± 0.46 per month, in which median is 1, range of minimum once a month to twice a month(Every 15 days). Compared with the previous study pattanashetti et al. is close to 1.09 ± 0.28 .

TABLE 3.6 Total number of transfusions done till the study done

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Total number of transfusions	140.2	103.7	132.0	6-480

Mean of total number of transfusions done up to the study period is 140.2 ± 103.7 , Median is 132.0 and from minimum number of transfusions is 6 to maximum 480 are transfused. We compare with the pattanashetti et al., means of the study is 151.40 ± 45.65 .

TABLE : 3.7 Hemoglobin level in the patients

Variable	Mean	Standard deviation	Median	Range (Min- Max)
Hemoglobin level (g/dl)	7.04	1.1	7	5-10

Mean of the hemoglobin level is 7.04 ± 1.1 g/dl,median is 7, Minimum is 5 mg/dl and maximum is 10g/dl. Compared with the previous study Md fazlul karim et al^[9]. Mean of their study is close to 7.2 ± 1.5 g /dl.

TABLE 3.8 : Level of MCHC in the patients

Variable	Mean	Standard deviation	Median	Range (Min- Max)
MCHC(g/dl)	36.48	1.54	34.10	32.2-37.6

Mean MCHC of the study is 36.48 ± 1.54 g/dl, median is 34.10 g/dl, and range is from 32.20 to 37.6 g/dl which is normal to higher side normal. Compared with the previous study Md fazlul karim et al. mean is 34.10 ± 2.8 g/dl.

TABLE 3.9 Gender distribution in the study

Gender distribution	Present Study
Male	29
Female	21
M:F ratio	1.38
Total	50

Total 50 patients were studied and among them 29 were male and 21 were female patients, with a ratio of 1.38 male to female. Comparison with the previous study, pattanashetti in the ratio was 4.83 among 35 patients with 29 male and 6 were females.

TABLE 3.10 Correlation with the total number of transfusions to the serum ferritin level

Number of blood Transfusion	Serum ferritin(Mean \pm SD)
<150	2948.09 \pm 1745.69
>150	3100.94 \pm 1365.42

Among total transfusions, patients who received more than 150 have more serum ferritin than the patients who received less than 150 units. Comparison with the study koreti s.^[9] in which mean in <150 units transfused 2109 ± 734 microgram /dl and in >150 units, 3956 ± 925 microgram/dl.

DISCUSSION:

Beta thalassemia is one of the common causes of hemolytic anemia in India. Here the hemolysis occurs due to defective hemoglobin synthesis and subsequently it leads to anemia. So it is very important to diagnosed earlier. Due to HbF symptoms are predominantly in 1 or after 1 year of age. In the present study the mean age of the diagnosis 19.22 ± 21.05 months (within 1 or less year) in the comparative Study with pattanashetti et al is 8.94 ± 6.15 months. Due to continuous hemolysis patient require the strict Monitoring of hemoglobin level for the further management and complications. In the present study the mean of hemoglobin level is 7.04 ± 1.1 g/dl which is close to the comparative Study (Md fazlul Karim et al) 7.2 ± 1.5 g/dl. So due to hemolysis these patients are required to frequently transfusion of blood. In our blood bank we issued Red cell concentrate. According to the hemoglobin level which is maintained in the body, some patients require it once or twice a month. In the present study among 50 patients, 68% are required once a month and 32% are required twice a month. Our age group is 1 month to 20 years, so from the age at which they were diagnosed and required transfusion up to the study done maximum 480 units were transfused. Due to repeatedly transfusion these patients are having complications like iron overload, allergic reactions, non-hemolytic reaction. So these patients are required to monitor with the serum ferritin level as it is easy to perform, low cost and had no side effects, other parameters can be monitored are liver biopsy, T2 based MRI assessment of liver and cardiac iron^[9]. In the present study the serum ferritin level in the patients of more than 150 units transfused is 3100.94 ± 1365.42 microgram/dl, while in less than 150 units 2948.06 ± 1745.69 microgram /dl. So here is the positive correlation with transfusion and mean Serum ferritin level. So it is mandatory to maintain serum ferritin level with chelation therapy. It will reduce the concentration of serum ferritin and it is effective in

preventing the iron induced tissue injury and prolonging life expectancy^[10]. In the present study 96% among the 50 patients are reliable on the chelation therapy, while only 4% are not required till the duration. The male gender is predominant in the beta thalassemia major in the present study 29 were male among the 50 patients, Male to female ratio is 1.38. The mean of the duration of the disease is 8.35 ± 4.57 years. The mean MCHC level is 36.48 ± 1.54 g/dl, which is upper side normal, compared with the previous study Md fazlul karim et al is 34.10 ± 2.8 g/dl. However in the anemia there is low MCHC level here due to spherocytes the MCHC level is normal or upper side normal.

IV. CONCLUSION:

Among the 50 patients of beta thalassemia major who are transfusion dependent mainly male predominant, with the mean age 9.48 ± 4.55 years having the 8.25 ± 4.57 years of duration of the disease, with the majority of patients required the once a month blood (red cell concentrate) to maintain Hemoglobin level is up to 10 gm/dl. The mean level is 7.04 ± 1.1 gm/dl. They required chelation therapy to maintain the serum ferritin level 1000 microgram/dl, it is the marker for the iron overload. So it is mandatory to monitor the duration of disease, hemoglobin level, serum ferritin level with the transfused units and frequency of transfusion in transfusion dependent beta thalassemia major patients, To give them better prolonging life expectancy and controlling the hemolysis and iron overload as well.

V. References

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