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To assess the serum ferritin level in multiple transfused patients of ß thalassemia major

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Abstract

Background: Globally, there are at least 60,000 individuals born with ß-thalassemia major (TM) each year[1]. Regular blood transfusions are mandatory for long term survival, but over a period of years these cause a secondary state of tissue iron overload.

Methods: Hospital based study was conducted on Patients of ß thalassemia major above 2 years of age received regular blood transfusions at least for 1 year duration, attending OPD in the Department of Pediatrics, S.P. Medical College, Bikaner.

Results: Maximum cases were from age group 3-8 years, where total 27 cases were observed and out of them 19, 7, 1 patients belonged to serum ferritin level (SFL) <2500, 2500-5000, and >5000 respectively followed by age group 9-12 years where total 14 patients were found and out of them 5, 8 and 1 patients belonged to serum ferritin level (SFL) <2500, 2500-5000 and >5000 respectively and minimum were from age group >12 years where only 9 patients were found and out of them 3 patients belonged to each group (SFL <2500, 2500-5000 and >5000).

Conclusion: There is an urgent need to rationalize the chelation therapy and to create awareness about the consequences of iron overload in the patients.

Keywords: SFL, TM, Chelation therapy.

Introduction

Globally, there are at least 60,000 individuals born with ß-thalassemia major (TM) each year[1]. Regular blood transfusions are mandatory for long term survival, but over a period of years these cause a secondary state of tissue iron overload. Myocardial iron deposition can result in cardiomyopathy, and heart failure remains the leading cause of death[2-4]. The introduction of the iron chelator deferoxamine greatly ameliorates the effects of iron toxicity, but long-term cardiac mortality has been very disappointing[3,5]. The ongoing deaths from cardiac iron loading may relate to inadequate compliance or genetic factors related to metal transporters not yet fully elucidated[6-8], but whatever the cause, there is strong evidence that long-term deferoxamine chelation does not effectively prevent myocardial siderosis in a majority of patients[9,10]. Deferiprone, the first approved oral chelator, has been shown in randomized controlled

trials to be effective monotherapy at 100 mg/kg/day dose in treating mild to moderately severe myocardial iron loading (myocardial T2* 8–20 ms), significantly improving both myocardial iron and ejection fraction[11], and the combination of deferiprone at 75 mg/kg/day with deferoxamine is likewise effective[12]. However greater total iron clearance is seen with combined treatment[13-15], which suggests that it might be useful for severe myocardial siderosis (T2* < 10 ms).

ß-thalassemia major is detectable in almost every Indian population group; however it is seen with higher frequency in north-west and Far East parts of the country[11]. Among different communities, Sindhis, Gujaratis, Bengalis, Punjabis and Muslims account for most cases of ß-thalassemia.

Materials And Methods

Study design: Hospital based cross sectional study.

Study duration: 12 months (August 2016 to July 2017).

Study place: Department of Pediatrics and in collaboration with department of Cardiology, S.P. Medical College and Associated Group of Hospitals, Bikaner

Study population: Patients of ß thalassemia major above 2 years of age receiving regular blood transfusions at least for 1 year duration.

Sample size: 50 cases meeting the criteria were included for the present study.

Sampling Method: Convenience sampling

Inclusion Criteria

Patients of ß thalassemia major above 2 years of age received regular blood transfusions at least for 1 year duration, attending OPD in the Department of Pediatrics, S.P. Medical College, Bikaner.

Exclusion Criteria:

1. Patients of ß-thalassemia with
2. Congenital Heart Disease
3. Ex-thalassemic patients undergone bone marrow transplant
4. Thalassemia intermedia
5. Patients on cardio toxic drugs

Data Collection

A detailed history and examination findings were noted in a predesigned proforma.

A detailed physical examination was done in each case and the vital parameters, signs of congestive cardiac failure and size of liver and spleen were noted. Chest X-ray, electrocardiogram (ECG) and 2-dimensional and doppler echocardiography were done.

Observations

Table 1: Distribution of Cases according to age group (years) in relation to serum ferritin level

Age Group (Years)	Serum Ferritin Level						Total	
	<2500		2500-5000		>5000			
	No.	%	No.	%	No.	%	No.	%
3-8	19	70.4	7	38.9	1	20.0	27	54.0
9-12	5	18.5	8	44.4	1	20.0	14	28.0
>12	3	11.1	3	16.7	3	60.0	9	18.0
Total	27		18		5		50	

Maximum cases were from age group 3-8 years, where total 27 cases were observed and out of them 19, 7, 1 patients belonged to serum ferritin level (SFL) <2500, 2500-5000, and >5000 respectively followed by age

group 9-12 years where total 14 patients were found and out of them 5, 8 and 1 patients belonged to serum ferritin level (SFL) <2500, 2500-5000 and >5000 respectively and minimum were from age group >12 years where only 9 patients were found and out of them 3 patients belonged to each group (SFL <2500, 2500-5000 and >5000).

Table 2: Distribution of Cases according to gender in relation to serum ferritin level

Gender	Serum Ferritin Level						Total	
	<2500		2500-5000		>5000			
	No.	%	No.	%	No.	%	No.	%
Female	9	33.3	10	55.6	0	-	19	38.0
Male	18	66.7	8	44.4	5	100.0	31	62.0
Total	27		18		5		50	

In present study, majority of patients were males 31(62%) while only 19(38%) were females. In serum ferritin level group <2500 total 27 patients were found and out of them 9 and 18 patients were females and males respectively, in SFL group 2500-5000 group total 18 patients were found and out of them 10(55.6%) were females and 8(44.4%) were males, while in SFL group >5000 total 5 patients were found and they all (100%) were males.

Table 3: Distribution of Cases according to Transfusion Requirement (ml/kg/year) in relation to serum ferritin level

Transfusion (ml/kg/year)	Serum Ferritin Level						Total	
	<2500		2500-5000		>5000			
	No.	%	No.	%	No.	%	No.	%
≤200	13	48.1	1	5.6	0	0	14	28.0
201-250	10	37.0	8	44.4	0	0	18	36.0
251-300	4	14.8	8	44.4	3	60.0	15	30.0
>300	0	0	1	5.6	2	40.0	3	6.0
Total	27	100	18	100	5	100	50	100
Mean	214.00		253.06		297.00			
SD	32.72		32.91		16.81			
f	18.308							
p	<0.001							

Mean blood transfusion requirement in SFL group <2500 was 214.00±32.72 ml/kg/year, in SFL group 2500-5000 was 253.06±32.91 and in SFL group >5000 was 297.00±16.81 ml/kg/year and this difference was found statistically highly significant (p<0.001).

Table 4: Distribution of Cases according to type of chelating agents in relation to serum ferritin level

Type of Chelating Agents	Serum Ferritin Level						Total	
	<2500		2500-5000		>5000			
	No.	%	No.	%	No.	%	No.	%
Deferasirox	27	100	17	94.4	5	100	49	98
Desferrioxamine	0	0	1	5.6	0	0	1	2.0
Total	27		18		5		50	
χ^2	1.814							
P	0.404 NS							

According to type of chelating agents received, in SFL group <2500, all 27(100%) patients had deferasirox type, in SFL group 2500-5000, out of total 18 patients 17(94.4%) and 1(5.6%) patient had deferasirox and desferrioxamine type respectively while SFL group >5000, all the patients had Deferasirox type of chelating agent and the difference was found statistically insignificant (p>0.05).

Discussion

In the present study, almost two third of the patients (66%), required annual blood transfusion of 200 to 300 ml/kg/year and compliance was satisfactory (>4days/week) in about half of the patients (50%). Serum ferritin was >2500 ng/ml in almost half of the patient (46%). Only 18% (9) of the patients were severely anemic.

In another study, done in India by Taksande et al, showed serum ferritin was >2500ng/ml in 65% of children, severe anemia in 67% and 24% children were on regular chelation therapy[16]. Pemde et al studied on 154 ß-thalassemia major patients and concluded that in spite of adequate chelation therapy, mean serum ferritin were approximately 3 times higher the desired value[17]Spirito et al in their study from Italy, on 32 ß thalassemia major patients age range from 12 years to 22 years, showed mean annual blood transfusion requirement was 180 ml to 591 ml. Mean serum ferritin level for their study was 2169 ng/ml[18]. Mostly patients (90%), were adherent to the chelation therapy. Riaz et al in their study on 79 ß Thalassemia patients with the mean age of 10.5 years, two fifth of patients not receiving iron chelation therapy at all[19].

Conclusion

There is an urgent need to rationalize the chelation therapy and to create awareness about the consequences of iron overload in the patients.

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