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Original research article

A study of splenectomy in beta: Thalassemia patients and its effects

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Abstract

Over thirty thousand children are born in India every year with this disorder.The two severe forms seen are thalassemia major(TM) and thalassemia intermedia (TI). Multiple long term effects due to chronic anaemia, tissue hypoxia and the compensatory reactions result in enhanced erythropoiesis and increased iron absorption. Additionally, regular blood transfusion leads to iron overload in critical organs such as the heart, liver, pancreas and gonads. If the annual red cell requirement exceeds 200 ml/Kg of RBC, splenectomy should be considered, provided that other reasons for increased consumption, such as hemolytic reactions, have been excluded. Other indications for splenectomy are symptoms of splenic enlargement, leukopenia and/or thrombocytopenia and increasing iron overload despite good chelation. This study is conducted to find the splenectomy as the treatment and its effects in the patients.

Keywords: Splenectomy, beta-thalassemia, paediatrics

Introduction

Beta-Thalassemia is a common inherited haemoglobin disorder resulting in chronic hemolytic anemia[1].Over thirty thousand children are born in India every year with this disorder[2].The two severe forms seen are thalassemia major(TM) and thalassemia intermedia (TI). Multiple long term effects due to chronic anaemia, tissue hypoxia and the compensatory reactions result in enhanced erythropoiesis and increased iron absorption. Additionally, regular blood transfusion leads to iron overload in critical organs such as the heart, liver, pancreas and gonads[3, 4, 5]. If the annual red cell requirement exceeds 200 ml/Kg of RBC, splenectomy should be considered, provided that other reasons for increased consumption, such as hemolytic reactions, have been excluded[6,7,8]. Other indications for splenectomy are symptoms of splenic enlargement, leukopenia and/or thrombocytopenia and increasing iron overload despite good chelation[9, 10].This cross sectional study would include children between 2-18 years of age, to be conducted over a period of 18 months. This study would be conducted in a tertiary care hospital for children.

Aims and Objectives

To study the Splenectomy in Beta-Thalassemia patients and its effects

Materials and Methods

Study design

This study was a cross sectional, observational study done in a tertiary care centre in Maharashtra. 100 thalassemic children above 2 years of age who have received at least 50 blood transfusions and/or have serum ferritin levels more than 2500 Ng/ml were studied. All children between 2-18 years of age who had fulfilled the inclusion criteria, were included in the study. Informed consent was obtained from all the parents of children and the proposal was approved by the institute ethics committee. The details were recorded in the case proforma designed for the study.

Sample size

According to test year data, approximately 100 Beta thalassemia patients who received more than 50 blood transfusion and serum ferritin level more than 2500 Ng/ml were treated in the hospital and hence sample size of present study will be approximately 100.

Study duration

July 2020 – Dec 2021.

Sampling method

Universal Sampling

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Inclusioncriteria

1. Children aged between 2-18 years.
2. Multiply transfused thalassemia children who have received more than 50 units of transfusions.

Exclusion criteria

1. Multiply transfused children with other hemolytic disorders.

Statistical analysis

Descriptive statistics of continuous variables were expressed as means and standard deviation. Discrete variables were presented as frequencies and group percentage. All continuous variables were tested for normal distribution by D'Agostino-Pearson normality test. Student's *t*-test was used to compare the means of continuous normally distributed data. Categorical data were tested using the chi-square test. All statistical tests were 2-tailed, and a p value of <0.05 was considered statistically significant. Data were analyzed using SPSS 17.0 software for Windows.

Results

Table 1: Age distribution in study subjects

Age (yrs.)	Percentage	No.
4 to 7	42.0%	42
8 to 11	40.0%	40
12 to 15	18.0%	18
Total	100.0%	100

Table 2: Gender distribution of study subjects

Sex	Percentage	No.
Female	38.0%	38
Male	62.0%	62
Total	100.0%	100

Table 3: Thalassemia major /Thalassemia intermedia vs serum ferritin

S. Ferritin Mean/Peak Recent		Thalassemia Major/Intermedia		Total
		Thalassemia Major	Thalassemia Intermediate	
2500 to 5000	No.	49	5	54
	%	53.8%	55.6%	54.0%
5000 to 7500 ^	No.	31	3	34
	%	34.1%	33.3%	34.0%
7500 to 10000 ^	No.	10	0	10
	%	11.0%	0.0%	10.0%
>= 10000 ^	No.	1	1	2
	%	1.1%	11.1%	2.0%
Total	No.	91	9	100
	%	100.0%	100.0%	100.0%

Table 4: Thalassemia major /Thalassemia intermedia vs mean Hb

Hemoglobin (gm%)		Thalassemia Major/Intermedia		Total
		Thalassemia Major	Thalassemia Intermediate	
< 8	No.	41	4	45
	%	45.1%	44.4%	45.0%
8 to 10 ^	No.	35	5	40
	%	38.5%	55.6%	40.0%
>= 10 ^	No.	15	0	15
	%	16.5%	0.0%	15.0%
Total	No.	91	9	100
	%	100.0%	100.0%	100.0%

Table 5: Percentage of splenectomy in study

Splenectomy	Percentage	No.
Yes	16.0%	16
No	84.0%	84
Total	100.0%	100

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Table 6: Splenectomy vs mean Hb

Hemoglobin (gm%)		Splenectomy done		Total
		Yes	No	
< 8	No.	10	35	45
	%	62.5%	41.7%	45.0%
8 to 10	No.	6	34	40
	%	37.5%	40.5%	40.0%
>= 10	No.	0	15	15
	%	0.0%	17.9%	15.0%
Total	No.	16	84	100
	%	100.0%	100.0%	100.0%

Table 7: Splenectomy vs. serum ferritin

S.Ferritin Mean/Peak Recent		Splenectomy done		Total
		Yes	No	
2500 to 5000	No.	7	47	54
	%	43.8%	56.0%	54.0%
5000 to 7500 ^	No.	4	30	34
	%	25.0%	35.7%	34.0%
7500 to 10000 ^	No.	5	5	10
	%	31.3%	6.0%	10.0%
>= 10000 ^	No.	0	2	2
	%	0.0%	2.4%	2.0%
Total	No.	16	84	100
	%	100.0%	100.0%	100.0%
Chi-Square tests	Value	DF	P-value	Association is-
Pearson Chi-Square $	9.807	3	0.02028	Significant
Pearson Chi-Square ^	0.389	1	0.533	Not significant

$ 3 cells (37.5%) have expected count less than 5. ^ Row data pooled and Chi-Square Test reapplied.
^ Continuity Correction applied.

Discussion

β-Thalassemia is an inherited hemoglobin disorder resulting in chronic haemolytic anemia that typically requires life-long transfusion therapy. Although traditionally prevalent in the Mediterranean basin, Middle East, India, and Southeast Asia has rendered β-thalassemia a global health problem.

In the present study, 100 thalassemic children above 2 years of age were studied. Mean age of the subjects studied was 8.5 years, the youngest being 4 years and oldest being 15 years of age. Maximum children belonged to the age group of 4-7 yrs.(42%), 40% belonged to age group of 8-11 yrs and 18% belonged to 12-15 yrs. 62% of study subjects were male and 38% were female. There was male preponderance in study. Similar study was done by SyarifRohimi*et al.* in March 2011 in which systolic and diastolic cardiac functions were studied in 34 regularly transfused thalassemic children. Mean age of subjects was 11.69 (SD 4.7 yrs., Range 2.6 to 20 yrs.)[11].

Out of 100 children studied, 91 children having thalassemia major and 9 of them had thalassemia intermedia. All of them were above 2 years of age who have received at least 50 blood transfusions and / or have serum ferritin levels more than 2500 Ng/ml as per inclusion criteria. Out of 91 children having thalassemia major, 49 (53.84%) had serum ferritin level between 2500-5000ng/ml, 31(34.1%) had serum ferritin level between 5000-7500ng/ml, 10(11%) had serum ferritin level between 7500-10000ng/ml and 1(1.1%) had serum ferritin level more than10000ng/ml. Similarly out of 9 children having thalassemia intermedia, (55.6%) had serum ferritin level between 2500-5000 Ng/ml, 3(33.3%) had serum ferritin level between 5000-7500 Ng/ml, and 1(1.1%) had serum ferritin level more than10000ng/ml. (table 6, graph 4). When Mean Hb was reviewed in 100 Thalassemic children, it was found that out of 91 children having thalassemia major,41 (45.1%) had mean Hb less than 8gm%, 35(38.5%) had mean Hb between 8-10gm%, 15(16.5%) had mean Hbmore than 10 gm%and out of 9 children having thalassemia intermedia, 4 (44.4%) had mean Hb less than 8 gm%, 5 (55.6%) had mean Hb between 8-10 gm%.

Out of 100 thalassemic children studied, 16 children were splenectomised. Indications of splenectomy in the transfusion-dependent patient was hypersplenism, increased blood transfusion frequency and for the adequate control of body iron with chelation therapy. Annual transfusion volume exceeding 225 to 250 mL/kg per year with packed red blood cell, may indicate the presence of hypersplenism. (The possible development of alloantibody should also be ruled out.) In present study, out of 16 splenectomised children, 10 were having Hb<8gm%; 6 were having Hb between 8-10 gm%. Also, after comparing with serum ferritin, incidence of splenectomy was 12.9% in age group having serum ferritin 2500-5000, 11.7% in age group having serum ferritin 5000-7500, and 50% in age group having serum ferritin 7500-

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10000. Percentage of splenectomy was more in age group having serum ferritin level between 7500-10000.

Finding were consistent with studydone by ZuhairOmranEasa*et al.*in 2009.This study was conducted on 140 patients with beta thalassemia major and thalassemia intermedia, expressed as two groups (group I) splenectomized patients and (group II) nonsplenectomized patients. In group I patients, 82.9% were under transfused and 80% were underchelated, whereas 91.4% of group II patients were under transfused and 74.3% of them were underchelated. Splenectomy was beneficial in reducing frequency of blood transfusion in77.1% of group I patients. Thus it can be concluded that, there was an aggravating effect of splenectomy on hemosiderosis. High serum ferritin level in splenectomised patients was associated with higher incidence of complications[12].

Conclusion

16 children were splenectomised. Serum ferritin level was higher in splenectomised group and ranged between 7500-10000 ng/ml.

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