P-ISSN: 2664-3685
E-ISSN: 2664-3693
www.paediatricjournal.com
IJPG 2018; 1(2): 32-35
Received: 06-09-2018
Accepted: 13-10-2018

Dr. Oniell Fernandes
Assistant Professor,
Department of Paediatrics,
Kanachur Institute of Medical Sciences, Mangalore,
Karnataka, India

Corresponding Author:
Dr. Oniell Fernandes
Assistant Professor,
Department of Paediatrics,
Kanachur Institute of Medical Sciences, Mangalore,
Karnataka, India

A study of splenectomy in beta-thalassemia patients and its effects

Dr. Oniell Fernandes

DOI: https://doi.org/10.33545/26643685.2018.v1.i2a.177

Abstract
Thalassaemia is a genetic disorder of the haemoglobin protein in red blood cells. It has been historically classified into thalassaemia minor, intermedia and major, depending on the genetic defect and severity of the disease. The clinical presentation of β-thalassaemia varies widely from a mild asymptomatic form in thalassaemia minor, to a severe disease in thalassaemia major where individuals are dependant on life-long blood transfusions. The hallmark of thalassaemia syndromes is the production of defective red blood cells that are removed by the spleen resulting in an enlarged hyperfunctioning spleen (splenomegaly). Removal of the spleen may thus prolong red blood cell survival by reducing the amount of red blood cells removed from circulation and may ultimately result in the reduced need for blood transfusions.

Keywords: Splenectomy, beta-thalassemia, paediatrics

Introduction
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.

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 the Department of Paediatrics, Kanachur Institute of Medical Sciences, Mangalore. 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 2017- June 2018

Sampling method: Universal Sampling.

Inclusion criteria
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

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 10: 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

Similar study was done by Syarif Rohimi *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 2500-5000 ng/ml, 31(34.1%) had serum ferritin level between 5000-7500 ng/ml, 10(11%) had serum ferritin level between 7500-10000 ng/ml and 1(1.1%) had serum ferritin level more than10000 ng/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 than 10000 ng/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 Hb more 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 %.

Finding were consistant with study done by Zuhair Omran Easa *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 in 77.1% 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.

References

1. Aessopos A, Farmakis D, Karagioraga M, *et al*. Cardiac involvement in thalassemia intermedia: a multicenter study. Blood. 2001;97:3411-341.
2. Sinha S, Black M, Agarwal S, *et al*. Profiling β-thalassaemia mutations in India at state and regional levels: implications for genetic education, screening and counselling programmes Hugo Journal. 2009 December;3(1-4):51-62.
3. Monte I, Capodanno D, Nicolosi E, *et al*. Atrial and ventricular function in thalassemic patients with supra-ventricular arrhythmias. Heart Int. 2009 June 30;4(1):e345-54.
4. Galanello R, Origa R, *et al*. Beta thalassemia, Galanello and Origa Orphanet Journal of Rare Diseases. Blood. 2010;5:11.
5. Thalassemia International Federation: Guidelines for the clinical management of thalassemia 2nd edition. 2008. [http://www.thalassemia.org.cy].
6. Borgna-Pignatti C, Galanello R, *et al*. Thalassemias and

related disorders: quantitative disorders of hemoglobin synthesis. Wintrobe's Clinical Hematology. 11th edition. 2004;42:1319-1365.
7. Borgna-Pignatti C, Vergine G, Lombardo T, *et al*. Hepatocellular carcinoma in the thalassemia syndromes. British Journal Haematology. 2004;124:114-117.
8. Galanello R, Piras S, Barella S, *et al*. Cholelithiasis and Gilbert's syndrome in homozygous beta thalassemia. British Journal Haematology. 2001;115:926-928.
9. Taher AT, Otrock ZK, Uthman I, *et al*. Thalassemia and hypercoagulability. Blood Review. 2008;22:283-292.
10. De Sanctis V, Tangerini A, Testa R, *et al*. Final height and endocrine function in Thalassemia Intermediary Journal Paediatrics Endocrinology, Metab. 1998;11:965-971.
11. Rohimi S, Advani N, Sastroasmoro S, *et al*. Tissue doppler imaging in thalassemia major patients: correlation between systolic and diastolic function with serum ferritin level Paediatric Indonesia. 2012;52:187-93.
12. Zuhair Omran Easa, *et al*. Complications of High Serum Ferritin Level after Splenectomy in â Thalassemic Patients. Kufa Med. Journal 2009;12(1):243-250.