



ROLE OF SURGICAL INTERVENTION IN HAEMOGLOBINOPATHY'S PATIENTS WITH SPLENOMEGALY

General Surgery

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ABSTRACT

Haemoglobinopathies like thalassemia and sickle cell anemia has various complications. Splenomegaly is one of these complications that increases morbidity. Aim of this study was to study the role and outcome of the splenectomy surgery in patients of haemoglobinopathies with splenomegaly. Fifty admitted patients of haemoglobinopathy with splenomegaly were operated for splenectomy and the haematological parameters like haemoglobin level, haematocrit value, platelets count and frequency of blood transfusion were studied before and six months after the surgery along with surgical complications. It was found that haemoglobin level, haematocrit value, platelets count were raised and the frequency of blood transfusion was reduced after splenectomy surgery. Complications of surgery observed were wound infection in 3 (75%) and pneumonia in 1 (25%) patient.

KEYWORDS

Sickle Cell Anemia, Thalassemia, Splenomegaly, Splenectomy

INTRODUCTION

Hemoglobin is produced by genes that control the expression of the hemoglobin protein and defects in these genes can produce abnormal hemoglobins which leads to various haemoglobinopathy like Sickle cell disease, Thalassemia, Hereditary spherocytosis, etc. Sickle cell disease (SCD) is an autosomal recessive disorder characterized by production of abnormal hemoglobin S and is associated with high morbidity and mortality¹. Thalassemia syndromes arise from deactivating, mutations or deletions in alpha or beta globin genes^{2,3}. In SCD chronic haemolysis leads to splenomegaly^{4,5,6,7}. In thalassemia, the ineffective RBC production, RBC production in spleen leads to splenomegaly. Splenomegaly is found to be associated with hypersplenism. If the splenomegaly causes an increase in frequency of blood transfusion, splenectomy may be required⁸. Present study was conducted to know the role and outcome of the splenectomy surgery for splenomegaly in patients of haemoglobinopathy.

AIM AND OBJECTIVES

To study the role and outcome of the splenectomy surgery in patients of haemoglobinopathies with splenomegaly.

Methodology

The present prospective study was conducted with all the patients of haemoglobinopathy with splenomegaly admitted at surgery wards of tertiary health care institute, of India from 1st January 2017 to 31st Dec 2017 and were followed up for a period of six months after treatment. With detailed history, clinical examination, blood investigations, abdominal ultrasound, diagnosis of complication of splenomegaly was done. Alkaline Haemoglobin Electrophoresis and High Performance Liquid Chromatography (HPLC)^{9,10} were done for diagnosis of haemoglobinopathies. The patients were admitted in wards and medical treatment was initiated. Patients of splenomegaly with hypersplenism underwent elective open or laparoscopic splenectomy. Blood transfusions were given as per requirement. All these patients were followed up for 6 months, with clinical examination, blood investigations, ultrasonography and assessed for the outcome of treatment.

Statistical Analysis:

Association between two continuous variables was analyzed by applying t test while that for categorical variables was calculated using Fishers' exact test. P value < 0.05 was considered to be statistically significant. Data was analyzed using SPSS software version 24. 55

OBSERVATIONS AND RESULTS

Table: 1 Age and Gender wise distribution of patients

Age (yrs)	Male		Female		Total	
	Number	Percentage	Number	Percentage	Number	Percentage
11-15	8	27.59	3	14.29	11	22
16-20	8	27.59	7	33.33	15	30
21-25	7	24.13	6	28.57	13	26
26-30	6	20.69	5	23.81	11	22
Total	29	100	21	100	50	100

Mean age \pm SD (years) = 20.2 \pm 5.09, Range: - 12-29 years

Mean age (Male) \pm SD (years) = 19.68 \pm 5.19, Range: - 12-29 years

Mean age (Female) \pm SD (years) = 20.90 \pm 4.97, Range: - 14-28 years

Table - 1 shows the age and gender wise distribution of patients. Age group represents the completed age in years. Majority of patients i.e. 15 (30%) were in the age group of 16-20 years. Mean age of patients was 20.2 years \pm 5.09 years. Amongst males majority of patients i.e. 8 (27.59%) were in the age group of 11-15 and 16-20 years. Mean age of patients amongst male was 19.68 \pm 5.19 years. Amongst females majority of patients i.e. 7 (33.33%) were in the age group of 16-20 years. Mean age of patients amongst females was 20.90 years \pm 4.97 years.

Table: 2 Haemoglobinopathy Pattern wise distributions of patients

Haemoglobinopathy	Pattern	Patients	
		Number	Percentage
Sickle Cell	AS	8	16
	SS	14	28
Thalassemia	Alpha	8	16
	Beta	20	40
Total		50	100

Table - 2 shows Haemoglobinopathy pattern wise distribution of patients. Majority of patients i.e. 20 (40%) had Beta- Thalassemia, followed by SS pattern of Sickle Cell Disease which is seen in 14 (28%) patients. Patients with Alpha - Thalassemia and AS pattern of Sickle Cell Disease each were 8 (16%) in number.

Table: 3 Distribution of haemoglobinopathy's patients according to surgeries performed for Splenomegaly

Surgery performed	Haemoglobinopathy's complication	Number	Percentage
Open Splenectomy	Splenomegaly with hypersplenism	16	32

Laparoscopic Splenectomy	Splenomegaly with hypersplenism	34	68
Total	-	50	100

Table - 3 shows the surgeries performed for splenomegaly in patients of haemoglobinopathy. Overall, 50 patients had underwent surgeries, out of whom 16 (32%) patients had open splenectomy and 34 (68%) patients had laparoscopic splenectomy.

Table: 4 Haematological Changes after Splenectomy in patients with Splenomegaly (n = 50)

Haematological parameters	Before Splenectomy Mean (SD)					After Splenectomy Mean (SD)					P Value
	Sickle cell		Thalassemia		Total	Sickle cell		Thalassemia		Total	
	AS	SS	Alpha	Beta		AS	SS	Alpha	Beta		
Mean Haemoglobin (gm/dl)	6.65 (0.27)	6.58 (0.28)	6.7 (0.44)	6.76 (0.35)	6.68 (0.33)	7.57 (0.42)	7.6 (0.49)	7.4 (0.37)	7.51 (0.50)	7.53 (0.46)	<0.0001
Mean Haematocrit (%)	21.71 (1.06)	21.32 (1.22)	22.03 (1.37)	22.1 (1.26)	21.81 (1.25)	28.2 (2.89)	27.96 (2.38)	28.13 (1.8)	27.83 (1.43)	27.97 (1.99)	<0.0001
Mean Platelets count (100000/ul)	1.22 (0.09)	1.19 (0.10)	1.27 (0.07)	1.28 (0.11)	1.24 (0.10)	1.87 (0.14)	1.83 (0.24)	1.9 (0.23)	1.96 (0.12)	1.9 (0.18)	<0.0001
Mean Frequency of blood transfusion (per 6 months)	2 (0.53)	2.35 (0.50)	2.25 (0.46)	2 (0)	2.14 (0.40)	1.37 (0.74)	1.28 (0.46)	1 (0)	1 (0.32)	1.14 (0.45)	<0.0001

Table - 4 shows the effect of splenectomy on haematological parameters. There were 50 patients that underwent splenectomy. 34 (68%) of them underwent laparoscopic splenectomy and 16 (32%) had open splenectomy. Mean haemoglobin level was raised from 6.68 (gm /dl) \pm 0.33 (gm /dl) to 7.53 (gm /dl) \pm 0.46 (gm /dl), mean haematocrit value was raised from 21.81 % \pm 1.25% to 27.97 % \pm 1.99%, mean platelets count was raised from 1.24 (*100000/ul) \pm 0.10 (*100000/ul) to 1.9 (*100000/ ul) \pm 0.18 (*100000/ ul) and mean frequency of blood transfusion per six months was reduced from 2.14 \pm 0.40 to 1.14 \pm 0.45 significantly with p value of <0.0001 after splenectomy surgery. After Splenectomy surgery mean haemoglobin level, mean haematocrit value, mean platelets count were raised and mean frequency of blood transfusion per six month was reduced significantly.

Table: 5 Complications after Splenectomy surgery in haemoglobinopathy's patients with splenomegaly (n = 50)

Surgery Performed	Complications			Percentage
	Post Operative Pneumonia	Wound Infection	Total	
Open Splenectomy (16)	0	2	2	12.5
Laparoscopic Splenectomy (34)	1	1	2	5.88
Total (50)	1	3	4	8

Table - 5 shows the various complications after splenectomy surgery in patients of haemoglobinopathy with splenomegaly. Overall, surgery was performed on 50 patients of haemoglobinopathy with splenomegaly. After surgery, complications were seen in 4 (8%) patients. Complication occurred was wound infection with open splenectomy surgery seen in 2 (12.5%) patients. With laparoscopic splenectomy, complication occurred in 2 (5.88%) patients, one had suffered from post operative pneumonia and other had wound infection.

DISCUSSION

The present prospective study was conducted in tertiary health care institute, of India, to know the role and outcome of splenectomy surgery in patients of Haemoglobinopathies with splenomegaly. The haematological parameters like haemoglobin level, haematocrit value, platelets count were found to be raised and frequency of blood transfusion required was reduced significantly in present study.

Al-Salem AH et al (1999)¹¹, Ammara SA et al (2014)¹², Mandal PK et al (2015)¹³, Chopra R et al (2005)¹⁴, Ghmaird A et al (2016)¹ and Porecha M et al (2009)¹⁵ had similar finding with present study of increased haemoglobin level after splenectomy in patients of haemoglobinopathy . Zhou YL et al (2014)¹⁶ also had consistent finding with present study. However, in their study they had compared the haemoglobin level of splenectomized patients with that of non splenectomized patients.

Al-Salem AH et al (1999)¹¹ found increase in haematocrit value after splenectomy. Zhou YL et al (2014)¹⁶ found increased reticulocyte count but this was not significant. This might be due to the fact that, they had not studied the haematocrit value in same group of patients before and after surgery but compared the reticulocyte count in two separate groups, one who underwent splenectomy and other who did not. Engluma BR et al (2016)¹⁷ did not get significant increase in reticulocyte count after splenectomy in patients of sickle cell disease.

This might be because they have done partial splenectomy in about 37% patients of splenomegaly, which did not result in more increase in reticulocyte count.

Al-Salem AH et al (1999)¹¹, Ammara SA et al (2014)¹², Zhou YL et al (2014)¹⁶ had similar findings, where the platelet count had significantly increased after splenectomy surgery. Chopra R et al (2005)¹⁴, Ghmaird A et al (2016)¹ had findings in line with present study for increased platelets count after splenectomy, however they had not tested it for significance.

Chopra R et al (2005)¹⁴, Mandal PK et al (2015)¹³, Porecha M et al (2009)¹⁵, Akhtar IK et al (2016)¹⁸, Ghmaird A et al (2016)¹ had findings in line with present study for reduction in requirement of blood transfusion after splenectomy surgery in different haemoglobinopathies, with different units of blood transfusion measurement and for different duration of follow up.

Complications observed in present study was wound infection in 3 (75%) and pneumonia in 1 (25%) patient. With open splenectomy surgery complication occurred was wound infection seen in 2 (12.5%) patients. With laparoscopic splenectomy surgery, complication occurred in 2 (5.88%) patients, one had post operative pneumonia and other had wound infection. There was no mortality after splenectomy.

Acute chest syndrome seen in 2 (1.39%) and Minor wound infection seen in 3 (2.09%) patients by Al-Salem AH et al (1999)¹¹, with morbidity seen in 5.6% (n= 143) of patients after splenectomy. Taher A et al (2006)¹⁹ found thromboembolic event in 4% of patients with thalassemia intermedia after splenectomy surgery. Porecha M et al (2009)¹⁵ had observed the complication as wound infection in 4 (8%) patients of thalassemia after splenectomy. Hassan MN et al (2011)²⁰, Ammara SA et al (2014)¹² observed a complication of portal vein thrombosis in 5 (3.12%) and in 1 (2.9%) patients of beta thalassemia after splenectomy, respectively. Mandal PK et al (2015)¹³ observed Post-splenectomy complications as: facial deformities in 21 (29.17 %), delayed pubertal growth in 17 (23.6 %), venous thromboembolism in 11 (15.28 %) and pulmonary hypertension in 5 (6.94 %) patients of beta thalassemia. These different complications were not found in present study. However, it was a retrospective study over three year period. The cause for the difference of complication in various studies after splenectomy could be due to different age group of patients at the time of surgery and different surgeons operating for population at different places, with haemoglobinopathy. However, the complication of post operative wound infection was seen in these studies was also found in present study.

CONCLUSION:

Splenectomy in haemoglobinopathy patients with splenomegaly improves the patient's haematological indices. Splenectomy in haemoglobinopathy patients with splenomegaly has more success rate with favourable outcome and less complication rate.

Conflict of Interest: None

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