



## International Journal of Surgery Science

E-ISSN: 2616-3470  
P-ISSN: 2616-3462  
© Surgery Science  
www.surgeryscience.com  
2019; 3(1): 04-06  
Received: 02-11-2018  
Accepted: 05-12-2018

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## Indications for splenectomy in a tertiary care hospital in South India

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**DOI:** <https://doi.org/10.33545/surgery.2019.v3.i1a.02>

### Abstract

**Background:** Splenectomy is a surgical treatment for a wide range of diseases including symptomatic splenomegaly, autoimmune, malignant, hereditary and congenital disorders, splenic injury/rupture secondary to blunt trauma to abdomen. By far the two most common atraumatic indications for splenectomy are malignancy and haematological autoimmune disorders, such as Idiopathic Thrombocytopenic Purpura (ITP) and Autoimmune Hemolytic Anemia (AIHA).

**Methods:** We did a retrospective study in a tertiary care hospital, of all patients undergoing splenectomy over a 5-year period to describe the indications for splenectomy.

**Results:** 186 patients underwent splenectomy, both emergency and elective (Open or Laparoscopic – conventional and hand-assisted). Most common indication was hematological in 153 patients, of which ITP-70 (37.6%) was the highest. Non-hematological indications were secondary to splenic cyst or abscess, neuroendocrine tumour of the pancreas, hydatid cyst of spleen. Emergency splenectomy was done for blunt trauma to abdomen, and grade 5 injury was commonly encountered. Among all, 16 (8.6%) patients had spleniculi.

**Conclusion:** In patients with hematological disorders, splenectomy must be undertaken only after anticipating both, short- and long-term risks and potential benefits to the patient.

**Keywords:** Idiopathic thrombocytopenic Purpura, auto-immune hemolytic anaemia, splenectomy, splenic trauma

### Introduction

Spleen mediates important immunologic, storage and hematologic function. A person can undergo splenectomy, not to cure the disease; but to alter the clinical course or providing symptomatic relief in disease. Currently, there are varied indications of splenectomy and it may benefit patients with an expanded spectrum benign and malignant diseases.

### Background

Condensation of the lymphoid tissue in dorsal mesogastrium, in the 5<sup>th</sup> week of embryogenesis, forms the spleen. The spleen starts off as a hematopoietic organ, and then later takes on the role of lymphoid organ in the 15<sup>th</sup> to 18<sup>th</sup> week of intra-uterine life. Normal spleen measures 12 x 7cm and is deficient of peritoneum in the hilar region <sup>[1, 2]</sup>. Spleen consists of the red and white pulp, which are incomplete parenchymal compartments, formed by intervening trabeculae. Central arteries within the white pulp, are surrounded by a sheath of T- lymphocytes and lymphoid follicles composed of B-lymphocytes. Red pulp contains sinusoids lined by fenestrated endothelium. The Antigen Presenting Cells (APCs), dendritic cells, are present between the red and white pulp <sup>[1]</sup>. It mediates immunological response to blood-borne antigens, by formation of antibodies and sequestration of antibody-coated blood cells, bacteria. <sup>[3,4]</sup> First splenectomy was done by Quittenbaum in 1826 <sup>[5]</sup>. Most common medical indication for splenectomy is a hematological disorder <sup>[6]</sup>. In most institutions, trauma is the primary indication for splenectomy, although it is becoming less common in recent years with more non-operative management of splenic injury <sup>[7]</sup>. Partial or complete splenectomy leaves the patient vulnerable to overwhelming infection by encapsulated bacteria such as Streptococcus pneumoniae, Haemophilus influenza, Neisseria meningitidis, which is known as Overwhelming Post-Splenectomy Infection (OPSI). Incidence of OPSI has reduced due to the widespread use of vaccinations and prophylactic use of antibiotics. Even then, there is a 5% lifetime risk of a serious infection <sup>[8]</sup> and the subsequent risk of mortality during one of these episodes is higher than expected in those with a spleen, at 40–54% <sup>[9]</sup>.

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In the current years, there is an increased risk of thrombosis, both venous and arterial, secondary to splenectomy [10]. Venous thromboembolism occurs in the splenoportal system, mostly in those with hereditary spherocytosis (HS), thalassaemia, other haemolytic anaemias, and trauma [11]. In thalassaemia and sickle cell disease, another vascular complication, pulmonary hypertension following splenectomy is known to occur, with high prevalence of 75% [12, 13]. The controversy that liver can perform this function in the absence of the spleen has been thought of. But it becomes important for the patient to have higher levels of specific antibody and an intact complement system for the same [14].

### Objectives

To describe the indications for splenectomy over a 5-year period in a tertiary care centre in South India.

### Methodology

For this purpose, we conducted a retrospective chart review in our hospital, including patients from October 2013 to October 2018. All patients who underwent splenectomy, both emergency and elective (Open or Laparoscopic – conventional and hand-assisted) were included for data collection. Those with incomplete data were excluded from the study. Details of a total of 186 patients was collected. Demographic details, such as age, gender, comorbidities, duration of disease condition, indications of splenectomy was collected. All statistical analyses were performed using SPSS V 16. Descriptive statistics such as frequencies, percentages, median and standard deviation was used to describe the demographic, clinical details and the indications. Institutional Ethical Clearance (Ref. No-220/2018) was obtained prior to starting the study.

### Results

Mean age of the patients was  $36.6 \pm 14.48$  years, of which 86 (46.2%) were females. A majority of the patients were in the age group of 18-45 years. (Table. 1) Most common indication was hematological in 153 patients, of which Idiopathic Thrombocytopenic Purpura (ITP) 70 (37.6%) topped the list. (Table. 2). Duration of disease condition ranged from 1-3 years. Those with ITP were refractory to steroids and hence, resorted to splenectomy. Hemolytic anemia included a maximum of patients with Auto Immune Hemolytic Anaemia (AIHA) 9(4.8%), with one patient having Evan's Syndrome. Non-hematological indications were for splenic abscess secondary to Bacterial endocarditis 1 (0.5%), hydatid cyst 5 (2.6%), pancreatic neuroendocrine tumour 2 (1%), splenic abscess 6(3.2%), splenic cyst 3 (1.6%). 141 (75.8%) had an elective splenectomy, conventional laparoscopic splenectomy or hand-assisted splenectomy. All elective patients had pre-operative vaccination as per protocol. There was a 3% conversion rate from laparoscopic to open splenectomy. 45 (24.1%) had emergency splenectomy, in view of blunt trauma to abdomen or spontaneous rupture of the spleen. Most commonly, Grade 5 injury, shattered spleen secondary to blunt trauma, was encountered. Among all, 16 (8.6%) patients had spleniculi, found near the splenic hilum and confirmed by histopathology. Remission of disease was attained in all, with only one patient relapsing secondary to enlarged accessory spleniculi, presenting as mass in left hypochondriac region. Splenic weight ranged from 20g to 3.2kgs, with the mean being  $505.8 \pm 566$  grams.

**Table 1:** Age distribution of patient undergoing splenectomy

Age Groups (Years)	No. (%)
18-45	102 (72.3%)
46-60	26 (18.4%)
>60	13 (9.2%)

**Table 2:** Hematological Indications for splenectomy

Indication	Number (%)
ITP	70 (37.6%)
Pancytopenia secondary to Hypersplenism	18 (9.6%)
Hereditary Spherocytosis	12 (6.4%)
Hemolytic Anemia	10 (5.3%)
Thalassemia	4 (2.1%)
Hodgkin's Lymphoma	4 (2.1%)
NHL	3 (1.6%)
Myelofibrosis	3 (1.6%)

### Discussion

A diseased spleen gives rise to two major syndromes – hypersplenism: anaemia (transfusion requirements exceeds 250 ml/kg of packed red blood cells per year), neutropenia (total leucocyte count <4,000/mm<sup>3</sup>) or thrombocytopenia (platelet count <1lakh/mm<sup>3</sup>) arising singly or in combination due to enlargement of spleen and symptomatic splenomegaly. [15] Reversal of pancytopenia or bicytopenia, confirms the diagnosis of hypersplenism [16]. Symptomatic splenomegaly is most commonly encountered in lymphoproliferative and myeloproliferative disorders. Splenectomy for myelofibrosis, is known not to prolong survival but indicated as a palliative measure for symptomatic splenomegaly. Second most common indication for splenectomy in our study was hypersplenism, 18 (9.6%). In patients with Idiopathic Thrombocytopenic Purpura (ITP), antibodies are formed to glycoproteins IIb/IIIa on the platelets. Spleen is the major site for formation of these auto-antibodies, as well as for platelet destruction. Small sized spleen and the presence of megakaryocytes in the bone marrow are diagnostic of ITP. Glucocorticoids are the main stay of treatment, followed by cytotoxic agents or immunoglobulins. 70 (39.6%) of our patients were steroid refractory, and hence underwent elective splenectomy. Musser *et al*, post-splenectomy, 77% of ITP patients show a complete response, 14% show a partial response, whereas only 9 % failed to demonstrate a significant improvement in platelet count [17]. In our study, 85% had a good response, 10% intermediate and 5% has relapse. Failure of splenectomy in these patients, is mainly attributed to accessory spleen or splenosis secondary to morcellation during surgery. Only one patient's relapse was attributed to presence of accessory spleen in our series. Thalassaemia syndrome includes a clinical spectrum of being asymptomatic like thalassaemia minor and intermedia to being transfusion dependent with iron overload states like thalassaemia major. There occurs a defect in one or more ( $\alpha$  or  $\beta$ ) globin chains of hemoglobin, resulting in ineffective erythropoiesis, haemolysis and consequent splenomegaly and hypersplenism. Splenectomy in these patients reduces the need for blood transfusion requirements [18]. 4(2.1%) patients had thalassaemia intermedia, with symptomatic splenomegaly. As already discussed, post-splenectomy, these patients are at a higher risk of thrombotic complications and sequelae, and hence, it becomes important to start anticoagulation therapy. AIHA, like ITP, it involves antibody-mediated cellular destruction and

complement activation within the splenic substance. In patients with Warm AIHA, favorable response to splenectomy occurs in 50-8-%<sup>[19]</sup>. Indication splenectomy in Hodgkin's 4 (2.1%) and Non-Hodgkin's 3 (1.6%), was required to make a positive diagnosis because of the suspicion of an underlying lymphoma, reticulosis or infiltrative condition. Several studies have found that post-splenectomy, these patients are at a risk of secondary leukemia, independent of treatment<sup>[20]</sup>. Pre-operatively, elective patients receive Pneumococcal and Haemophilus influenza vaccination, atleast 2 weeks prior to the surgery. In case of emergency cases, they receive vaccination after 2 weeks.

### Conclusions

The spleen, both anatomically and physiologically in diseased condition, may significantly worsen the clinical picture in a variety of medical disorders such as ITP, Pancytopenias and hemolytic anemias. And hence, splenectomy must be undertaken only after anticipating both, short- and long-term risks and potential benefits to the patient. It should also be kept in mind about the ability of an asplenic individual to mount an adequate antibody response, relates to the indication, age and the degree of underlying immune suppression. Even with effective prophylactic measures to prevent OPSI and thromboembolic events, it becomes necessary to follow up these patients at regular intervals and, for vaccination.

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