



# Long-term Outcome of Splenectomy in Immune Thrombocytopenia (ITP)

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## **Author's contribution**

*The sole author designed, analyzed, interpreted and prepared the manuscript.*

## **Article Information**

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## **ABSTRACT**

ITP is a chronic illness with frequent mucosal bleeding. Though the first line treatment is corticosteroids, relapse is frequent which necessitates splenectomy. Response to splenectomy is usually good initially; however, over the years relapses are frequent. Drugs then used are azathioprine, mycophenolate mofetil, anti-D globulin, TPO-receptor agonists, rituximab, cyclosporine, etc. Ultimately there is a small subset of patients who may require a small dose of prednisolone lifelong.

**Keywords:** *Splenectomy; mycophenolate mofetil; rituximab; cyclosporine.*

## **1. INTRODUCTION**

The standard of care for individuals with steroid-refractory ITP for many years was splenectomy. However, the situation has altered due to the rituximab and TPO-RAs (Thrombopoietin Receptor Agonists) availability.

The first-line treatment for ITP is corticosteroids. Normally, a small dose of prednisolone is found good enough to give an adequate response to the patient.

However, after about a year of response, the platelet count drops again and the patient may

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have spontaneous mucosal bleeding like gum bleed, epistaxis, etc. Few female patients may have increased menstrual bleeding. All these progresses suggest a patient may develop refractory ITP. Splenectomy should be taken into account in this case due to the spleen's tendency for excessive platelet destruction.

### 1.1 Splenectomy Efficiency for ITP

Platelet counts rise right post-splenectomy, and patients experience long-lasting remission in the majority of ITP patients [1-3]. In a study of published case series from 1966 to 2004, 1731 (66 percent) of 2623 individuals kept a full response after a median of twenty-nine months of follow-up [4].

Twenty to thirty percent of individuals experience post-splenectomy relapses during the first 24 months [5]. However, they respond to alternative treatment. A small proportion of patients experience high mortality and morbidity [5].

The International Working Group Consensus survey [7] and ASH ("American Society of Hematology") in 2011 [6] both propose splenectomy as 2<sup>nd</sup> line treatment for ITP. However, Splenectomy is more strongly suggested by ASH guidelines.

### 1.2 Predictors of Splenectomy Response

Splenectomy outcomes are unpredictable, and data on response rates pertain to people instead of specific patients. Splenectomy response predictors were assessed as follows below: It has been suggested that the initial week following splenectomy might indicate long-term response.

Complications connected with splenectomy might be infections. It is widely known that asplenic patients run the risk of developing severe bacterial sepsis [7]. This risk was decreased from about 7.16/100 to 2.3/100 patient years with pre-splenectomy vaccination [8].

### 1.3 Platelet Scintigraphy

Indium-labeled "autologous platelet scanning", in which the patient receives [9] In-labeled autologous platelets and following scintigraphy determines the location of platelet clearance [10], has become an effective predictor of "splenectomy" response but was not yet attained broad recognition. A splenectomy response was

seen in 91.4 percent (range between 87.0-94.8 percent) of people with a "splenic pattern" and 40.9 percent (range between 15.4-100 percent) of people having a diffuse, mixed, or hepatic pattern within a pooled study of 6 cases (n=580) [11,12].

## 2. PONDICHERRY EXPERIENCE, INDIA [13]

### 2.1 Aim

Splenectomy is the recommended 2<sup>nd</sup> line therapy for chronic immunological thrombocytopenic purpura after corticosteroids fail to control the condition. Splenectomy has a 50–60 percent success rate in obtaining complete remission. However, the real long-term result is uncertain because the majority of follow-ups are less than five years, as per cited articles. This retrospective analysis of adult people having chronic ITP evaluates the genuine long-term result of splenectomy across a long time and the determinants of splenectomy response.

### 2.2 Methods

We studied the medical records of 49 patients at a reputed institution in southern India who just had a splenectomy for chronic ITP between 1982 and 2015 [Range between 2 to 396 months (33 years)]. The average duration of observation was 75.71 months.

### 2.3 Results

Splenectomy response decreased from 83.3 percent (5 out of 6 patients) after 1 year to 25 percent (1 out of four patients) when evaluated after fifteen years. There was no statistically substantial correlation between any of the examined variables, including age, gender, the first reaction to steroids ("pre-operative"), long-term remission, as well as post-operative platelet count. Nevertheless, Individuals having "post-splenectomy" recurrence may be treated with a much lower dosage of prednisolone (16.7 mg per day) than their typical pre-splenectomy demand (47.5mg per day).

### 2.4 Conclusion

Even though splenectomy temporarily "normalizes platelet count", maintained long-term remission gradually declines with time; just one-third of patients may still be in remission by the

ten years following surgery. Furthermore, no element could predict how a splenectomy would respond.

A review by Vesely SK et al. [14] showed that 656 splenectomized patients treated with 22 treatments were examined in a systematic review by Vesely SK et al. The greatest complete responses were recorded with azathioprine, cyclophosphamide, and rituximab, although only in 41-109 individuals. The reported full response rates varied from 17-27%, although 36-42 percent of patients with such three therapies did not respond. Most publications solely discussed platelet count responses; only 63 patients' bleeding outcomes were mentioned (10 percent). The only individuals with a pretreatment platelet count less than  $10 \times 10^9$  cells/L were 111 (17%) of the 656 eligible patients.

Bell et al. [15] found that there is insufficient data to help the efficacy of any therapy for individuals with chronic severe thrombocytopenia and ITP following splenectomy. Randomized, controlled trials should be used to assess potentially beneficial therapies for both benefit and safety.

### **3. LONG-TERM EFFECTS OF ITP SPLENECTOMY**

ITP is predominantly an acquired thrombocytopenia-related condition that does not include marrow failure. Splenectomy was initially utilized in 1913 to treat ITP. With the discovery that opsonin (crucial for the optimum elimination of invading microorganisms with white blood cells is produced within the spleen, spontaneous splenic excision has been questioned and reassessed. The success rate of Splenectomy is approximately constant (approximately 50 and 60 percent), regardless of whether it is carried out very early after diagnosis or several months or years afterward.

### **4. PERSPECTIVE: DECIDING ON SPLENECTOMY**

In general, it is advised splenectomy should be deferred at least for a year after the onset of illness, since there may be spontaneous remission.

There are several situations in which splenectomy should be considered. Patients may have severe ITP, which is characterized by severe bleeding, profound "thrombocytopenia", a poor or temporary response to IVIg and

corticosteroids, as well as a suboptimal reaction to TPO-RA. Splenectomy is performed for express relief.

## **5. CONCLUSION**

ITP is a benign recurrent bleeding disorder that responds to steroids initially very well. However, relapses are common which warrant consideration of splenectomy later. Nevertheless, the long-term effect of splenectomy might not be very satisfying in all cases; and patients may require subsequent treatment with oral drugs like rituximab, TPO-R agonists, cyclosporine, low dose steroids, etc.

## **CONSENT AND ETHICAL APPROVAL**

It is not applicable.

## **COMPETING INTERESTS**

Author has declared that no competing interests exist.

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