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Splenectomy for β-Thalassemia Major Among Transfusion-Dependent Patients: Effectiveness and Complication

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ABSTRACT

Objectives: To assess the effectiveness and complications of splenectomy in thalassemia major patients dependent on repeated blood transfusions.

Methodology: This retrospective descriptive study was conducted in the Paediatric and Neonatal Surgery Department of a tertiary care hospital from January 2020 to January 2024. The study included 43 β-thalassemia patients (median age: 13 years; range: 9–17 years) who required frequent blood transfusions from thalassemia units.

Preoperative ultrasonography was performed in all patients to detect accessory spleens. Post-splenectomy outcomes were evaluated, including Changes in annual blood transfusion requirements, Platelet level variations (pre- and post-splenectomy), Quality of life (parent-reported) and Complications (intra-abdominal hemorrhage, wound site bleeding, respiratory infections)

Results: Blood transfusion needs decreased significantly post-splenectomy. A notable rise in platelet levels was observed after splenectomy. Serum ferritin levels declined significantly, indicating reduced iron overload. Parents reported improved quality of life in their children. 3 patients had incision site bleeding, managed with diathermy coagulation and blood/platelet transfusion. 3 patients developed fever three months post-splenectomy, requiring medical intervention.

Conclusion: Splenectomy in thalassemia major patients resulted in a sustained reduction in transfusion needs, increased platelet counts, and enhanced quality of life. However, any subsequent rise in transfusion requirements should prompt further investigation to identify potential underlying causes.

Keywords: Thalassemia, Post-splenectomy complications, Blood transfusion

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Introduction

Thalassemia is a group of inherited disorders resulted from absent of globin chains either alpha or beta hence resulting in α-thalassemia or β-thalassemia.[1] β-globin chains defect resulting in β-thalassemia is considered to involve approximately 1.5 % of the global population.[2] This results in defective red blood cell (RBC) morphology, and hence the lifespan of RBC is reduced.[3] In thalassemia patients, splenomegaly occurs due to increased red blood cell destruction and extra medullary haematopoiesis. Splenectomy (removal of the spleen) may improve red blood cell survival by reducing the number of cells destroyed and potentially decreasing the need for blood transfusions.[4] Initially the thalassemia was classified according to the mutation in gene and nature of the disease into thalassemia minor, thalassemia inter media and thalassemia major. Now a days it is classified

according to the need of blood transfusion as mild, non-transfusion-dependent thalassemia (NTDT) and as severe, transfusion-dependent thalassemia (TDT).[5] The major management for, transfusion-dependent thalassemia (TDT) is to regularly transfuse red blood cells(RBC) and the resultant increase In iron overload is prevented by iron chelation therapy i.e. deferasirox (DFX), desferrioxamine (DFO)[6] . Currently, the only known cure for this condition is a transplant of stem cells from a matching donor,i.e allogeneic stem cell transplantation.[7] One of the function of the spleen is to remove abnormal red blood cells from the circulation. Splenomegaly leads to reduction of red blood cells, white blood cells (WBCs) and platelets in the blood stream ad also enlarged spleen causes discomfort and pain to the patient. Thus, removing spleen may be of benefit in treating patients with TDT.[3,8]Splenectomy is often necessary for patients with Transfusion-Dependent Thalassemia (TDT) due to hypersplenism, characterized by Increased red blood cell transfusion requirements (>200-220 mL/kg/year), symptomatic splenomegaly and severe pancytopenia. The Thalassemia International Federation (TIF) also recommends removal of spleen for Increased blood transfusions requirement that cannot be managed by chelation therapy having hypersplenism and symptomatic splenomegaly. [9]

Methodology

This retrospective descriptive study was conducted at the Department of Paediatric and Neonatal Surgery from January 2020 to January 2024, involving 43 patients with transfusion-dependent thalassemia (TDT) aged 9-17 years. We collected detailed clinical histories focusing on quality of life indicators, documented infectious episodes and hospitalizations, and analyzed complete blood counts before and after splenectomy, including hemoglobin levels, platelet counts, and white blood cell counts. Additional parameters evaluated were blood transfusion requirements and serum ferritin levels both pre- and post-operatively. Post-splenectomy outcomes were carefully monitored, with particular attention to surgical complications such as wound site bleeding and intra-abdominal hemorrhage, infectious complications, and mortality.

Results

During the study period, 43 thalassemia patients underwent splenectomy with a mean age of 13 years (range 9-17 years), comprising 25 female and 18 male patients. All patients received vaccinations against meningococcus, Haemophilus influenzae type b, and pneumococcus 4-6 weeks preoperatively (Table I).

Preoperative hematological parameters revealed a mean hemoglobin level of 7.6 ± 0.7 g/dL, which was optimized through preoperative blood transfusions. The mean total leukocyte count (TLC) was 7315 ± 3131/mm³, while mean platelet count was 178 ± 112 × 10³/mm³ prior to surgery. Postoperative follow-up demonstrated significant improvements in both hemoglobin and platelet values compared to presplenectomy levels (P<0.001).

The mean presplenectomy serum ferritin level was 3202 ± 1497 ng/mL, which showed significant reduction during follow-up (P<0.001). Annual blood transfusion requirements decreased markedly from 290 ± 35 mL/kg/year presplenectomy to 201 ± 23 mL/kg/year in the first postoperative year, and further to 160 ± 23 mL/kg/year at final follow-up (P<0.001) (Table II).

Table I: Demographic and Clinical Characteristics of β-Thalassemia Major Patients Undergoing Splenectomy.

Parameter	Value
Number of patients (n)	43
Age (years)	13 (average: 9-17)
Sex	
Male	25
Female	18
Accessory Spleen	3
Cholilithiasis	3
Concurrent cholecystectomy	1
Family history	3
Vaccination	43
Complications (n=39)	
Bleeding	4
Infection	3
Mortality	1

Concurrent cholelithiasis was identified in 3 patients (7%), with cholecystectomy performed in 1 case. Accessory spleens were detected and removed in 3 patients (7%). Postoperative complications included

Table II: Presplenectomy / Postsplenectomy Comparison of Blood Count and Need of Blood Transfusion

Parameter	Presplenectomy	Postsplenectomy	p-value
Hemoglobin level (g/dl)	7.6 ± 0.7 g/dl	9.8 ± 0.7 g/dl	< 0.001
White blood cell count (/mm³)	7315 ± 3131	11735 ± 2741	< 0.001
Platelet count (150,000-400,000/mm³)	178,000 ± 112,000	253,000 ± 123,000	< 0.001
Serum ferritin (ng/ml)	3202 ± 1497 ng/ml	2229 ± 1405 ng/ml	< 0.001
Blood transfusion requirement (ml/kg/year)	290 ± 35 ml/kg/year	201 ± 23 ml/kg/year	< 0.001

incision site bleeding in 4 patients (9.3%) managed with fresh frozen plasma and platelet transfusions, and surgical site infections in 3 patients (7%). One mortality (2.3%) occurred due to sepsis. Parental reports indicated substantial improvement in quality of life following the procedure.

Discussion

Splenectomy is primarily indicated for transfusion-dependent thalassemia (TDT) patients to reduce both splenic blood consumption and transfusion requirements, while also mitigating iron overload.[2] However, current guidelines recommend a cautious approach to splenectomy due to the significant risk of overwhelming post-splenectomy infection (OPSI), particularly restricting the procedure for children under 5 years of age. [3,10] In our study cohort of 43 patients, the mean age was 13 years (range 8-17 years), with all patients receiving appropriate vaccinations against pneumococcal, Haemophilus influenzae type b, and meningococcal infections 4-6 weeks preoperatively.

The threshold for defining "high transfusion requirement" justifying splenectomy varies across guidelines, ranging from 200-275 mL/kg/year.[8,10,11]. Our patients demonstrated a mean preoperative transfusion need of 290±35 mL/kg/year. The hematologic response to splenectomy in β-thalassemia patients shows considerable variability across studies. Merchant et al. [18] reported a reduction from 294.85±22.6 mL/kg/year to 138.41±90.38 mL/kg/year post-splenectomy, while Muhammad et al.[7] observed a decrease from 219.6 mL/kg/year to 125.5 mL/kg/year (p=0.001). These transfusion reductions significantly improve patients' quality of life, as demonstrated by Zhou et al.[12] Our findings align with these studies, showing substantial decreases in transfusion requirements postoperatively.

The hematologic benefits extend beyond transfusion needs. We observed significant postoperative increases in platelet counts, consistent with Muhammad et al.'s findings[7], along with notable reductions in serum ferritin levels, mirroring the trends reported by Zhou et al.[12]

Preoperative evaluation should include assessment for cholelithiasis, particularly in symptomatic patients, with consideration for concurrent cholecystectomy when indicated,[13] Akca et al.[1] identified gallstones in 32.4% of cases, performing cholecystectomy in only 36.3% of these patients. Similarly, we detected gallstones in 3 patients (7%), with cholecystectomy performed in just one case due to the asymptomatic nature in others.

Accessory spleens (splenunculi), occurring in 10-30% of the general population [5,14], may contribute to persistent transfusion requirements post-splenectomy.[15,16] Akca et al.[1] reported accessory spleens in 32.4% of cases, with one diagnosis made eight years post-splenectomy due to increasing transfusion needs. Our protocol includes routine preoperative ultrasonographic screening for accessory spleens, which were identified and removed in 3 patients (7%) during the primary procedure. This proactive approach helps minimize the risk of recurrent symptoms and maintains reduced transfusion dependence.

Conclusion

Splenectomy in transfusion-dependent thalassemia patients significantly reduces blood transfusion requirements, increases platelet counts, and improves quality of life. However, any subsequent rise in transfusion needs should prompt investigation for potential causes, such as accessory spleen or alloimmunization. Close postoperative monitoring is essential to ensure sustained benefits and detect complications early.

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