Original Article

Splenectomy in Children with Beta Thalassemia Major and Sickle Cell Disease: A 4-Year Experience from Iraq

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Abstract

Background: Splenectomy is indicated as a therapeutic option in different types of hereditary blood disorders. **Objective:** The aim of this study is to determine the outcome of splenectomy in children with beta-thalassemia major (BTM) and sickle cell disease (SCD). **Materials and Methods:** A retrospective study was conducted in Basrah-Iraq on children with BTM and SCD who had undergone open splenectomy in the period between January 2016 and December 2019 at our institution. **Results:** The study included 87 children (49 males and 38 females). 59.8% with SCD and 35 (40.2%) with BTM. The mean age was 8.46 range (3–15) years. The mean weight was 23.51 range (11–45) kg. In SCD, the major indication (71.1%) was acute splenic sequestration (ASS). Postsplenectomy outcome in BTM included improvement in the median hemoglobin (Hb) level by 2.1 g/dL, reduction in the median packed red blood cells (PRBC) transfusion frequency by 19/year, and reduction in the median PRBC requirement by 190 mL/kg/year. In SCD, improvement in the median Hb level by 1.55 g/dL, reduction in the median PRBC transfusion frequency by 4.5/year, and reduction in the median PRBC requirement by 45 mL/kg/year. In addition, splenectomy eliminated life-threatening ASS crisis. Postoperative complications were observed in 6.9% of patients. **Conclusions:** SCD was the main hematological condition requiring splenectomy. ASS was the main indication for splenectomy in SCD. Splenectomy has proved beneficial in the management of SCD and BTM patients. Furthermore, it is now recognized as a safe surgery with a low complication rate.

Keywords: Children, sickle cell disease, splenectomy, splenic sequestration, thalassemia, transfusion

Introduction

Splenectomy is indicated as a therapeutic option in different types of hereditary blood disorders. Hemoglobinopathies are prevalent hereditary diseases in the Middle East area including Iraq.[1-3]

Regarding thalassemia; splenectomy considered under three clinical scenarios: increased blood requirement that prevents adequate control with iron chelation therapy, hypersplenism, and symptomatic splenomegaly.[4,5] Splenectomy is indicated in patients with sickle cell diseases (SCD) having recurrent acute splenic sequestration (ASS) crisis or after a single life-threatening attack.[6]

Many clinical and hematologic consequences are anticipated after splenectomy including an increase in baseline hemoglobin (Hb), decrease blood transfusion requirement, increase in platelet and white blood cells count.[7,8]

Regardless the indications; splenectomy has its possible complications, especially sepsis, and thromboembolic phenomena.[4,6]

The aim of this study is to determine the outcome of splenectomy in children with beta-thalassemia major (BTM) and SCD.

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Materials and Methods

A retrospective cross-sectional study was conducted in Basrah, Iraq. The records of children with BTM and SCD who had undergone open splenectomy in the period between January 2016 and December 2019 at our institution were reviewed. Ten patients with other hereditary blood disorders were excluded [thalassemia intermedia (n = 6), hereditary spherocytosis (n = 3), and one patient with congenital dyserythropoietic anemia]. Additionally, patients who underwent laparoscopic splenectomy were not included. Furthermore, one patient who died at zero postsplenectomy day excluded as well. Variables collected were the age at splenectomy, weight, sex, hematological disorder, indication for splenectomy, postoperative complications, residency, in addition to the pre- and post-splenectomy hematological and clinical parameters including the Hb level, packed red blood cells (PRBC) transfusion frequency and the PRBC transfusion requirement.

The diagnosis of hematological disorders was established in the hematology department by clinical features and confirmed by investigations including Hb electrophoresis. Indication for splenectomy according to our institution's guideline were mainly hypersplenism, one major or two minor ASS attacks, increase requirements of PRBC transfusions more than 250 mL/kg/year.

The criteria required to determine hypersplenism included splenomegaly with any combination of anemia, leukopenia, thrombocytopenia, in association with hypercellular bone marrow. In patients with SCD, ASS was divided into minor attacks in which there a fall in Hb level of 3 g/dL below the basal level of the patient, and major attacks in which there is a drop in Hb level by more than 4 g/dL.

All patients examined preoperatively by ultrasound for the presence of gallstones. Additionally, patients received vaccination against postsplenectomy infections at least 2 weeks before surgery. Preoperatively, PRBC and platelet concentrates were infused when required, Hb was maintained above 9 gm/dL.

Splenectomy performed under general anesthesia by open surgical technique via Left subcostal incision. Patients were covered with intravenous third-generation cephalosporin for at least 3 days postoperatively, Nasogastric tube kept for at least 24 h. On discharge, long-term penicillin prophylaxis was advised.

The outcome was examined by the capability of splenectomy to decrease the PRBC transfusion requirements in patients with thalassemia and to remove the risk of splenic sequestration crisis and improve Hb level in SCD patients. Hb level recorded preoperatively before any blood transfusion and postoperatively as a mean of six readings during the year after surgery.

The study approved by the institutional review board of our institution. Written informed consent was obtained from patients' families.

Data were analyzed with SPSS version 22. A P value less than 0.05 was considered as statistically significant. for comparison between pre- and post-operative variables, Wilcoxon signed-rank test was used.

Results

This study involved 87 children, 35 (40.2%) with BTM and 52 (59.8%) with SCD who were undergone splenectomy in a public pediatric hospital in Basrah; a city in southern Iraq. The subdivision of SCD were sickle cell anemia (n = 31), sickle beta zero thalassemia (n = 7), and sickle beta plus thalassemia (n = 14).

The mean age of the study population was 8.46 range (3–15) years. The mean weight was 23.51 range (11–45) kg. Males were more than half the patients (49, 56.3%). Most of the patients were from the peripheries of Basrah (72.4%).

In BTM, the indication for splenectomy was increased PRBC transfusion requirement more than 250 mL/kg/year, 31.4% (11/35 thalassemic patients) had hypersplenism as well. In SCD, the most common indication for splenectomy was splenic sequestration in 37/52 (71.2%), the remaining were due to hypersplenism in 15/52 (28.9%) [Table 1]. Seven (8%) patients underwent simultaneous cholecystectomy for gallstones.

In patients with BTM, there was postsplenectomy improvement in the median Hb level by 2.1 g/dL, reduction in the median transfusion frequency by 19/year, and reduction in the median PRBC requirement by 190 mL/kg/year. Similar improvements were also noted in patients with SCD: improvement in the median Hb level by 1.55 g/dL, reduction in the median transfusion frequency by 4.5/year, and reduction in the median PRBC requirement by 45 mL/kg/year [Table 2]. Furthermore, splenectomy eliminated life-threatening ASS crisis attacks.

Postoperative complications were observed in 6 (6.9%) patients. Among them, three (two BTM and one SCD) experienced pneumonia and atelectasis, one SCD patient had a surgical site infection, one BTM patient developed acute gastric dilatation, and one SCD patient developed acute chest syndrome [Table 3].

Discussion

Data from 87 splenectomized patients have been analyzed. The mean age of the patients who underwent splenectomy was 8.46 years which is older than in the Egyptian series 6.68 ± 2.54 years,[9] Although the population is relatively

Table 1: Demographic characteristics of the study populations ($n = 87$)

Parameter	BTM ($n = 35$)	SCD ($n = 52$)
Sex (male/female)	18/17	31/21
Age in year, median (IQR)	10 (7–12)	8 (6–10)
Body weight in kg mean (minimum–maximum)	23.43 (11–42)	23.58 (13–45)
Residency: (center/periphery)	7/28	17/35
Indications for splenectomy		
Increased transfusion requirement	24 (68.6%)	0
ASS	0	37 (71.2%)
Hypersplenism[a]	11 (31.4%)	15 (28.8%)

BTM: beta thalassemia major, SCD: sickle cell disease

[a]Hypersplenism cases had frequent blood transfusion as well

Table 2: Response to splenectomy according to the type of hematological disorder

Hematological disorder	Transfusion parameter	Presplenectomy, (median, IQR)	Postsplenectomy (median, IQR)	*P* value[a]
BTM ($N = 35$)	Hemoglobin (g/dL)	6.1 (5.9–7.1)	8.2 (7.9–8.7)	<0.0001
	Transfusion frequency	25 (23–31)	6 (5–8)	<0.0001
	PRBC requirement (mL/kg/year)	250 (230–310)	60 (50–80)	<0.0001
SCD ($N = 52$)	Hemoglobin (g/dL)	6.45 (5.9–7.2)	8 (7.5–8.7)	<0.0001
	Transfusion frequency	5.5 (4–22.8)	1 (1–4)	<0.0001
	PRBC requirement (mL/kg/year)	55 (40–190)	10 (10–40)	<0.0001

BTM: beta thalassemia major, IQR: interquartile range, SCD: sickle cell disease

[a]Wilcoxon signed-rank test

Table 3: Postoperative complications

Complication	BTM ($N = 35$) N (%)	SCD ($N = 52$) N (%)	Total ($N = 87$) N (%)
Pneumonia and atelectasis	2 (5.7)	1 (1.9)	3 (3.45)
Surgical site infection	0	1 (1.9)	1 (1.1)
Acute gastric dilatation	1 (2.9)	0	1 (1.1)
Acute chest syndrome	0	1 (1.9)	1 (1.1)
Total	3 (8.6)	3 (5.8)	6 (6.9)

BTM: beta thalassemia major, SCD: sickle cell disease

younger than in the Tabuk series in which the mean age of 34 patients was 20.7 ± 6.6 years old,[10] this diversity is greatly attributed to the heterogeneity in the population studied that included both thalassemia and SCD patients. A study in Oman revealed age results similar to our study.[11]

Splenectomy in thalassemia is of benefit in decreasing blood transfusion requirement and relieving patients from the consequences of hypersplenism and physical discomfort resulting from massive splenomegaly.[12,13] Regarding patients with SCD, previous reports revealed that splenectomy can of benefit in case of ASS or hypersplenism.[14]

In our series, SCD and BTM were the most common indications for splenectomy. SCD represents the majority of cases (59.8%) because of 68.38% of our registered patients were SCA till the end 2019.

In total, the indications for splenectomy in this study were Increased transfusion requirement in 24, ASS in 37, and hypersplenism in 26 patients (11 thalassemia and 15 SCD). About 42.5% of all splenectomies were due to ASS. Regardless the age among SCD, the majority of indications were ASS (71.2%). This finding was nearly similar to the experience from Oman (60.4%) of splenectomized patients in SCD due to ASS.[11] Similar findings documented in a study conducted in Saudi Arabia in which ASS was the commonest indication for splenectomy in 103 (76.9%) patients.[15]

The other indications for splenectomy in patients with SCD was hypersplenism (28.9%), this finding is less than Oman study (36.4%)[11] and higher than Saudi Arabia study (13.4),[15] but nearly similar to experience from Oman (mean 6.25 years).[11]

In patients with BTM, an appropriate transfusion therapy is necessary to suppress endogenous erythropoiesis and to prevent bone deformity and cardiomegaly.[16] Splenomegaly is known to develop in BTM patients, which causes red blood cells destruction and increases the

need for blood transfusions. Consequently, splenectomy is advantageous for these patients.[17-19]

In our BTM patients, after splenectomy, PRBC requirements significantly reduced to 60 mL/kg/year (76% reduction), median PRBC transfusion frequency significantly reduced from 25 to 6 per year, median Hb level significantly increase from 6.1 to 8.2 g/dL. Nearly similar results reported by other studies.[20,21] splenectomy can decrease blood transfusion to one per month in Memon *et al*.[22] instead 4–5 time and to half in Akhtar *et al*.[23]

Splenectomy in SCD patients prevent further ASS, additionally, it has been reported to decrease the transfusion requirement by 45% for 6–12 months.[24] Furthermore, a study in Tabuk (Saudi Arabia) reported that following splenectomy for SCD resulting in decreased given blood from 4.5 to zero per year, and the median Hb level improved from 9 ± 1 g/dl to 11 ± 2 g/L postoperatively.[25]

In our SCD patients, splenectomy eliminated ASS and reduced requirement of given blood to 10 mL/kg/year in 81.8% resulting in transfused of one time per year with increased Hb from 6.45 to 8 g/dL. Although there was study done appeared increased in Hb level to 3.4 g/dL,[11] while in another study there was no significant increase in Hb level.[7]

We reported six (6.9%) postoperative complications, 8.6% of the BTM and 5.8% of the SCD patients, which is similar to another two studies done in Pakistan,[22,26] and higher than the result done,[10] the difference in the results may be due to the difference in the sample size (only 34 patients included in the comparative study).

In SCD patients, 8.4% postsplenectomy complications were reported by a study in Tabuk.[25] Older publications documented postsplenectomy complications that ranged from 13% to 49%.[27-29] These figures have greatly improved as highlighted in recent papers and in our experience due to advancements in surgical and anesthetic techniques as well as a better understanding of the pathophysiology of various hematological disorders.[24,30,31]

In children with SCD, acute chest syndrome is a significant cause of mortality and morbidity.[32] Kalpatthi *et al*.[33] reported postoperative acute chest syndrome in 6.9% of 58 SCD patients underwent splenectomy for ASS. In our series acute chest syndrome observed in one (1.9%) of 52 SCD patients.

The limitations of the present study might have affected the results. First, there was no comparison with the patients who did not undergo splenectomy. Additionally, due to an insufficient registry, we did not analyze crucial factors including immunological characteristics and iron overload markers.

Conclusions

SCD was the most common hematological condition requiring splenectomy in Basrah, Iraq society. ASS was the main indication for splenectomy in SCD which was observed in older patients (around 7 years old) when compared with other publications. In BTM, the major indication for splenectomy was increase PRBC transfusion requirement. In general, splenectomy has proved beneficial in the management of SCD and BTM patients. In patients with SCD, it prevented ASS and improved Hb level significantly, whereas in BTM patients, it reduced the median transfusion requirement to 60 mL/kg/year. Splenectomy is a safe surgery with a low complication rate.

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Conflict of interest

There are no conflicts of interest.

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