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# Acute chest syndrome after splenectomy in children with sickle cell disease

Salim Ghantous, Saad Al Mulhim\*, Nafeesa Al Faris, Basel Abushullaih, Firas Shalak, Salam Yazbeck

Saudi Aramco Medical Services Organization, Dhahran 31311, Saudi Arabia

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Sickle cell disease; Acute chest syndrome; Splenectomy; Children

#### Abstract

**Background/Purpose:** The purpose of this study was to determine the incidence of acute chest syndrome (ACS) in children with sickle cell disease (SCD) undergoing laparoscopic or open splenectomy and to assess factors that may predispose to this complication.

**Methods:** A retrospective review of all patients with SCD undergoing splenectomy between 1999 and 2007 in our institution. Charts were screened for demographics, perioperative clinical status (vaso-occlusive crises, sequestration crises), preoperative hemoglobin electrophoresis and preoperative transfusion, postoperative development of ACS, and need for an intensive care unit (ICU) admission. **Results:** Forty-three children with SCD, 17 females and 16 males (mean age 9 years), underwent splenectomy (19 laparoscopy and 24 open). Acute chest syndrome occurred in 9 patients (20%), 1 (5.2%) of 19 in the laparoscopy group, and 8 (33.3%) of the 24 in the open group. All patients with ACS were admitted to the ICU. Acute chest syndrome developed within the first 24 hours in 5 of the 9 patients, on the second postoperative day in 1 patient, and more than 1 month postoperatively in 3 patients. Six of 9 patients with ACS had been transfused preoperatively. All patients with ACS had had vaso-occlusive crises before surgery. Five of 9 patients who developed ACS had previous ACS episodes before surgery. There was no death in our series.

**Conclusion:** The incidence of ACS is in accordance with the literature. Preoperative transfusions did not prevent ACS. There is a clear tendency for laparoscopically operated patients to experience less ACS postoperatively. In our group of patients, there were no clear benefits for routine perioperative admission to the ICU.

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Acute chest syndrome (ACS) is the most serious cause of hospitalization and intensive care unit (ICU) admissions among patients with sickle cell disease (SCD). Children with SCD undergoing splenectomy are at increased risk for developing ACS because of their decreased ability to defend against infection, as well as postoperative pain and complications of general anesthesia [1]. In the pediatric population with SCD, there is an increase risk for postoperative complications resulting from surgical stress and from traumatic large incisions previously required to remove enlarged spleens. Laparoscopic splenectomy with intracorporeal fragmentation is thought to reduce the incidence of

Dhahran 31311, Saudi Arabia. *E-mail address:* saad.mulhim@aramco.com (S. Al Mulhim).

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\* Corresponding author. Surgical Division, SAMSO, PO Box 76,

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postoperative ACS in children. Our aim with this study was to determine the incidence of ACS in children with SCD undergoing laparoscopic and open splenectomy and to assess factors that may predispose to this complication.

### 1. Materials and methods

A retrospective review of all patients with SCD under the age of 16 who underwent splenectomy between 1999 and 2007 in our institution was conducted, and charts were screened for demographics, perioperative clinical status (vaso-occlusive crisis and sequestration crises), perioperative hemoglobin electrophoresis and perioperative transfusion, postoperative development of ACS, and need for an ICU admission.

Acute chest syndrome was defined as the combination of one or more of the following signs and symptoms: hypoxemia with  $Sao_2$  less than 90%, chest pain, dyspnea, tachypnea, cough radiologic evidence of bronchopulmonary infiltrates, raised white cell count, and fever higher than  $38.5^{\circ}$ C [1-3].

#### 2. Results

A total of 43 children with SCD, 17 females and 26 males, with a mean age of 9 years, underwent splenectomy: 19 laparoscopic and 24 open (Table 1).

The median follow-up was 3.5 years (0-7 years).

Acute chest syndrome occurred in 9 patients (20%): 1 of 19 in the laparoscopy group and 8 of 24 in the open surgery group.

All patients with ACS were admitted to the ICU. Acute chest syndrome developed in 5 of the 9 patients in the first 24 hours, on the second postoperative day in 1 patient, and more than 1 month postoperatively in 3 patients. Six of 9 patients with ACS had been transfused in the immediate preoperative period. There was no death in our series and none required ventilatory support.

Of the whole series, 28 patients were transfused in the immediate preoperative period, of whom 6 had ACS. Of the 15 who were not transfused 3 developed ACS.

 Table 1
 Comparison of laparoscopic and open splenectomy groups

	Open surgery	Laparoscopic surgery
Age (years)	9.79	7.57
Sex ratio (M/F)	10/14	7/12
Mean preoperative hemoglobin level (g/L)	10.6	9.9
Mean operating time (min)	104.91	153.15
Postoperative length of stay (d), median (range)	9 (5-21)	5 (3-16)

	Postoperative ACS (n = 9)	No postoperative ACS (n = 34)
Age (y)	9.55	8.58
Sex (M/F)	7/2	18/16
Preoperative hemoglobin level (g/dL)	9.8	10.2
Laparoscopic approach	1	18
Open surgery	8	16
Mean operating time (min)	126.77	123.67
Postoperative hospital stay (d), median (range)	10 (5-16)	5 (3-14)

Three patients with ACS had concomitant surgeries, whereas this was the case for only 9 of the 34 patients who did not develop ACS.

### 2.1. Comparison of laparoscopic and open splenectomy patients

Table 1 compares perioperative data between patients who underwent laparoscopic and open splenectomy. The average age in the open group was higher than in the laparoscopy group (9.8 vs 7.6 years). Younger age patients are considered to be a higher risk group for developing ACS.

The operative time was longer in the laparoscopic group: 153 vs 105 minutes (P = 0.6%).

Postoperative hospital stay was longer in the open group than in the laparoscopy group ( 9 vs 5 days).

## 2.2. Comparison of ACS patients to non-ACS patients after splenectomy

Acute chest syndrome occurred in 9 (20%) of 43 patients. All patients with ACS were admitted to the ICU (Table 2).

Acute chest syndrome developed in the first postoperative day in 5 of the 9 patients, on the second postoperative day in 1 patient, and more than 1 month after surgery in 3 patients. Two of those 3 were known to have previous episodes of ACS before splenectomy. Six of the 9 patients with ACS had been transfused in the immediate preoperative period.

Although all patients in the study previously had vasoocclusive crises, there was a clear tendency for the laparoscopically operated patients to develop postoperative ACS less frequently (1/19) than those undergoing an open splenectomy (8/24).

Postoperative hospital stay was prolonged if ACS occurred (10 vs 5 days) (P = .03).

### 2.3. Children who developed ACS

Acute chest syndrome occurred in 2 females and 7 males. Ages ranged from 3 to 14 years. Preoperative hemoglobin level ranged between 8.7 and 11.9 g/dL (Table 2).

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