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Predictors of mortality in adults with Sickle cell disease admitted to intensive care unit in Bahrain☆,☆☆

Sana Abdulaaziz Al Khawaja [a,*], Zainab Mahdi Ateya [b], Ridha Abdulla Al Hammam [b]

[a] Salmaniya Medical Complex, Intensive Care Unit, P.O. Box: 12, Bahrain
[b] Intensive Care Unit, Salmaniya Medical Complex, Ministry of Health, Intensive Care Unit, P.O. Box: 12, Bahrain

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ABSTRACT

Objective: Sickle cell disease (SCD) is one of the most common genetic blood disorders in Bahrain. However, there is a paucity of data regarding the clinical presentation of SCD patients who require ICU admission. This study aimed to describe the epidemiological data of SCD patients admitted to the ICU and to identify predictors of mortality in order to help intensivists identify patients at most risk.
Method: This study used a retrospective, descriptive, and correlational design. Records of 210 SCD patients admitted to the ICU in Salmaniya Medical Complex (SMC) were reviewed.
Results: 210 patient with SCD were admitted to ICU during study period. The main reason for ICU admission was acute chest syndrome. The mortality rate of SCD patients in the ICU was 12.7%. The four significant predictors of mortality were age, number of previous hospitalizations, length of stay, and need of renal replacement therapy. Non-survivors were older than survivors (M = 38.6 vs. M = 30.3 years, U = 1685, z = −2.2, P = 0.025), were less frequently hospitalized (M = 19 vs. M = 46 times, U = 1274, z = −3.7, P = 0.000), had shorter lengths of stay in the ICU (M = 3.1 vs. M = 5.5 days, U = 1145, z = −4, P = 0.000), and more likely to require renal replacement therapy the p = 0.0297 than the survivors. The combined effect of these predictors was modelled with cox regression and found to be statistically significant. Gender, presence of comorbid conditions, level of hemoglobin on admission, and need for blood transfusions were not found to be predictors for mortality.
Conclusion: Acute chest syndrome was the main reason for SCD patients to be admitted to the ICU. Older age, less frequent hospitalization, shorter stays in the ICU, and the need for renal replacement therapy were found to be indicators of high mortality rate SCD patients.

1. Introduction

Sickle cell disease (SCD) encompasses a group of inherited hemoglobin disorders caused by mutation in the beta-globin chain of the hemoglobin molecule. This result in the formation of sickle hemoglobin (HbS), which has the singular property of polymerizing when deoxygenated [1]. Polymerization of deoxygenated sickle hemoglobin leads to distorted sickle shape of red blood cell (RBC) with shortened lifespan [2]. Through a complex interplay of adhesive events among blood cells, these altered erythrocytes can obstruct the vasculature, producing episodes of pain, hemolytic anemia, organ injury, and early mortality [3]. Sickle patients are vulnerable to multiple ICU admissions secondary to theses complication [2]. Although the molecular basis of SCD is well characterized, the complex mechanisms underlying vaso-occlusion (VOC) have not been fully elucidated [3].

Sickle cell disease is the most prevalent hereditary hematological disease worldwide [4]. SCD causes significant morbidity and mortality, particularly in people of African and Mediterranean ancestry. In a large study in Bahrain, were a 56,198 blood samples of Bahraini nationals were analyzed over a six year period. The prevalence of SCD was reported as 2.1%. Al-Arrayed and coworkers reported that the mild form of the disease predominates in Bahrain, with a wide clinical variability [3].

The majority of SCD patients receiving their care and follow up in the Salmaniya Medical Complex (SMC), which is the biggest and the main governmental hospital. A special SCD unit has been developed in SMC for the management of various complication associated with this disease. This unit deals mainly with the stable chronic complication and management of simple vaso-occlusive and hemolytic crises. If these patients develop acute complications, become haemodynamidcally unstable, or need organ support they will be shifted to the ICU.

Since SCD affects mainly young population, and death among these patients are often sudden, it has a large social and cultural impact in our country [6,7-11].

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* Corresponding author.
E-mail addresses: SKhawaja3@health.gov.bh (S. Al Khawaja), Zateya@health.gov.bh (Z.M. Ateya), rhammam@outlook.com (R.A. Al Hammam).

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2. Method

2.1. Setting

The study was conducted at the Salmaniya Medical Complex, which is the main governmental hospital in the Kingdom of Bahrain.

2.2. Sample/population

All sickle cell patients (n = 210) admitted to the ICU between January 2011 and December 2012 were included in the study. Their records were obtained from the ICU patient registration book.

2.3. Design

This study is quantitative, descriptive, correlational, and cross sectional in design.

2.4. Eligibility and exclusion criteria

All sickle cell patients admitted to the unit in the specified period were included regardless of their age, gender, nationality, or number of admissions.

2.5. Data collection instrument

A data collection form was developed by the research team. The form includes the patient's demography, hospitalization information, reason for admission, comorbidities, supportive treatments, laboratory findings upon ICU admission, and the final outcome upon ICU discharge. The tool was piloted to ensure appropriateness; some amendments were made before the final usage.

2.6. Data collection procedure

The major sources of the patients' data were the ICU patients' registration book and the individual patient files as acquired from the filing section. Meticulous care was taken with identification of data and data entry was double checked to ensure accuracy.

2.7. Data analysis

Gathered data was analyzed using IBM SPSS 20. Descriptive and inferential statistics for the demographic variables, illness-related variables, mortality rate, and the predictive model were calculated with a confidence interval of 95%. Then the relationships between these variables were explored with the appropriate statistical procedures depending on the distribution of variables' values. Since the mortality predictors were bond with time to event, Cox regression model was created to illustrate this relationship.

3. Results

During the study period, 210 patients with SCD were admitted to the ICU, accounting for 24% of the total ICU admissions during this period. Of these admissions, 117 (56%) were female patients. The range of patients' ages at admission was 14 to 65 years, with a mean of 31 years, compared to a range of 14 to 97 years with a mean of 47 for all ICU admissions. The majority of the patients, 133 (%63) were received in the ICU from the general ward after a few days stay, and the remaining (37%) were admitted directly from the accident and emergency department. The average length of stay for SCD patients was 5.4 days, compared to 5.1 days for all other ICU admissions.

The reasons for admission to ICU were grouped under three main categories: medical (directly related to SCD complications); surgical (included trauma and planned post-operative treatment); and obstetric (included pregnant sickle cell disease patient with complications during their pregnancy requiring ICU admission, in addition to planned post caesarian section admission; Table 1). It was difficult to choose a single reason for ICU admission for some of our patients, as some patients may have a complex clinical presentation with more than one complaint and more than one organ involved. This complexity may be related to the nature of SCD and the sickling process. The most frequent medical reason for admission to ICU was acute chest syndrome (ACS) (n = 115, 55%). The six gastrointestinal reasons for admissions included three with obstructive jaundice, for which they needed endoscopic retrograde cholangiopancreatography (ERCP) for diagnostic and therapeutic purposes, two cases of sickle cell disease hepatopathy with a bilirubin level > 500 mmol/L, and one case of upper gastrointestinal bleeding secondary to esophageal varices in a cirrhotic patient. The two neurological cases included one patient with ischemic stroke and another patient with status epilepticus. The three cases of sepsis were admitted to ICU with multiple organ failure secondary to bacteremia. All obstetrics patients survived their ICU stay.

The ten surgical reasons for ICU admission included three patients with acute cholecystitis, two with intestinal obstruction, and one with pancreatitis. All of them were managed conservatively. One case of acute mesenteric ischemia, which required explorative laparotomy, and three planned post-elective surgical admissions (one post splectomy, two post laparoscopic cholecystectomy). There were no cases of trauma requiring ICU admission.

Upon admission to ICU, 11% of SCD patients had co-morbidities, including pulmonary embolism, lower limb deep venous thrombosis, chronic renal failure, ischemic heart disease, diabetes mellitus, hypertension, hepatitis, chronic liver disease, obstructive sleep apnea, epilepsy, pulmonary hypertension, systemic lupus erythematosus, and stroke.

We did not have enough information about the prophylactic use of hydroxycarbamide among our study population.

Of the 210 admissions, twenty six died in the ICU, a mortality rate of 12.7%, compared to an overall mortality of 17.2% for all adults admitted to the ICU.

We found that the mortality rate greatly varied across the year, to create two prominent seasons of higher mortality. The first season is two months long, encompassing January and February, during this period, the mortality rate rises to 11.5%. The second season of death among SCD patients runs from June until the end of October, the mortality rate reaches 15.7% during this period (Fig. 1).

Table 1
Causes of intensive care unit admission for sickle cell patients.

Cause of admission	Number N(210)	Percentage
Medical	**187**	**89**
Acute chest syndrome	115	54.7
Hemolytic crises	30	14
Vaso-occlusive crises	14	6.6
Pulmonary embolism	8	3.8
Gastrointestinal	6	2.8
Transfusion related lung injury	6	2.8
Sepsis	3	1.4
Aplastic crises	2	0.9
Neurological	2	0.9
Splenic sequestration	1	0.4
Obstetric	**13**	**6.2**
Acute chest syndrome	4	1.9
Vaso-occlusive crises	4	1.9
Planned post-caesarean section	3	1.4
Hemolytic crises	2	0.9
Surgical	**10**	**4.8**
Acute cholecystitis	3	1.4
Planned post-operatively	3	1.4
Intestinal obstruction	2	0.9
Pancreatitis	1	0.4
Acute mesenteric ischemia	1	0.4