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Fever in sickle cell disease patients in the Kingdom of Bahrain

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Summary Sickle cell disease (SCD) is a hereditary blood disorder characterized by abnormal red blood cell shape (sickling). The incidence of the disease in the Kingdom of Bahrain is approximately 2.1%. Patients with SCD are prone to multiple health complications, some of which are fatal. A retrospective study was conducted at the Salmaniya Medical Complex in the Kingdom of Bahrain from June, 2012 to December, 2012 to assess the incidence of fever among the SCD in-patients at that institution. The study also assessed the antibiotics administered, type of organisms isolated and patient outcome. The results showed that a total of 59 patients developed fever and 33 of those with fever had a positive culture result (55.93%). The most common isolate was gram-positive bacteria, most commonly *Staphylococcus epidermidis* (42.86%), and ceftriaxone was the most commonly prescribed antimicrobial. In conclusion, there was a low rate of hospital acquired infection and special attention needs to be paid to the infection control measures for SCD patients. For most of the SCD fever cases, there was no clear cause of the fever even after extensive diagnostic evaluations; thus, those fevers may have been of a non-infectious etiology in this population. Fortunately, none of the patients had any of the common preventable infections as a result of the high vaccination coverage and strong vaccination program in place in the Kingdom of Bahrain.

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Introduction

Sickle cell disease (SCD) is a hereditary blood disorder characterized by abnormal red blood cell shape (sickling). This abnormal shape decreases the cell's flexibility and results in multiple complications. This abnormality is due to a mutation in the hemoglobin gene that results in an amino acid substitution of valine for glutamine in the β chain. The mutation can occur in a heterozygous (SCD trait) or homozygous form. The disease has multiple acute and chronic complications, including hemolysis crises, severe pain, renal complications, thromboembolic phenomenon and overwhelming infections, and some complications of SCD generate high mortality.

Infections in SCD patients are relatively common and are spread easily for various reasons. SCD patients are prone to invasive infections with encapsulated bacteria. SCD patients should be assumed to be functionally asplenic even if their spleen is not surgically removed. Bacteremia carries significant risk of morbidity and mortality in this group of patients despite the protocols for routine vaccination.

In a study conducted in a hospital population in Bahrain, which included 5503 neonates and 50,695 non-neonates, the prevalence of SCD was reported as 2.1 percent. The prevalence of the HbS trait was reported as 18.1 percent in neonates, and only 10.44 percent of the SCD patients were non-neonatal patients [1]. Thus, SCD patients occupy a significant percentage of the Bahraini population and, as a result, impose a considerable burden on Bahraini health services. The natural progression of the disease among those patients, in particular the prevalence of infection, has not been studied previously. This study explored this part of the disease to raise more awareness of its significance on morbidity and mortality.

Most studies on this medical topic involved children. A study performed at the University of California about the predictors of bacteremia in SCD children found that white cell count, absolute neutrophil count and level of bandemia were all independently associated with bacteremia [2]. Another study in Jamaica conducted from 1993 to 1996 investigated the cause and outcome of fever in Jamaicans with homozygous SCD. The study concluded that acute chest syndrome and painful crises were the most important complications associated with fever in SCD patients [3].

As mentioned above, little research has been done on adults with SCD. Therefore, we aimed to explore the cause and development of fever in adult patients with homozygous SCD during their hospital stay. We also examined the outcome of those patients and the prevalence of different bacterial infections/hospital acquired infections in SCD patients. This help reveal the prevalence of infections in this population and whether any infection prevention methods can be implemented on clinical grounds.

Methods

This was a retrospective study conducted at the Salmaniya Medical Complex, the main secondary care and teaching hospital in the Kingdom of Bahrain. The rate of sickle cell disease patients in that hospital on a daily basis is approximately 10%. At the same time, 20—30% of the accident and emergency visits at that hospital are for sickle cell disease patients.

Data were collected retrospectively from all of the medical records of patients who were admitted to the medical department with a diagnosis ICD 10 of sickle cell disease from June, 2012 to December, 2012.

The medical records of the SCD in-patients that had a fever at their time of admission or developed a fever during their hospitalization were evaluated. Then, all cases that developed a healthcare-associated infection (HAI) is a localized or systemic condition resulting from an adverse reaction to the presence of an infectious agent(s) or its toxin(s) that was not present on admission to the acute care facility.

The information for the study forms was collected from the medical record files by medical residents who were involved in the care of these patients. The information collected included: demographic data, the chief complaint of the patient at admission, admission diagnosis, onset and duration of fever, clinical symptoms and signs, type of antimicrobials used and the duration of their use, laboratory blood investigation on admission and follow up, hospital stay events, whether subspecialty consultation was necessary, radiology investigations performed and the impression at discharge. The data were reviewed and analyzed by an infectious disease specialist.

Results

There were a total of 565 admissions during the period of June, 2012 to December, 2012. Most of these patients had been repeatedly admitted