

## SICKLE CELL DISEASE: PATIENTS' AWARENESS AND MANAGEMENT

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Sickle cell disease (SCD) is a genetically transmitted disorder of red blood cells which is characterized by severe hemolysis and recurrent vaso-occlusive episodes. Another important feature of the disease is patients' increased susceptibility to acute viral and bacterial infections.<sup>1</sup>

The sickle cell gene is frequent in almost all the Arabian Gulf countries,<sup>2,3</sup> and the reported incidence of sickle cell trait in Bahrain is about 11%,<sup>4</sup> with nearly 50 homozygous cases being added every year. A consanguinity rate of about 43% among Bahrainis is perhaps one of the many reasons for the persistence of the sickle cell gene in this population.

SCD commonly manifests itself as a painful crisis affecting joints and limbs, and factors such as infection, dehydration, exhaustion, and a change in temperature may precipitate this crisis. There is a wide variation in the severity of the pain, from mild transient attacks lasting only a few hours to severe pain of longer duration, with some patients even requiring hospitalization. In Bahrain, since hospital facilities are easily available and patient care is free of charge, SCD patients through the Accident and Emergency Units for the relief of even minor pains. Some genuine cases, however, require hospital admission, as these cannot be managed at home. Since SCD is associated with increased morbidity and mortality, it is important for patients to have some awareness of its causes and the genetic implications. A pilot study of SCD patients admitted to Salmaniya Medical Center was carried out. The objectives were to assess the patients' knowledge about SCD, as well as their own level of education, and to review the mode of management.

### Materials and Methods

This cross-sectional pilot study included 118 Bahraini SCD patients (85M, 33F) admitted to Salmaniya Medical Center over a four-month period from May to August

1995. In order to assess the patients' health knowledge and educational background, all the SCD patients were interviewed personally, and their responses to preset questions were recorded. A second form was devised to include presenting clinical manifestations and management. For this information, the patient's medical charts were reviewed. To determine the statistical significance, unpaired *t*-tests were performed using the INSTAT program (San Diego, CA, Graph PAD software).

### Results

The age of the 118 Bahraini SCD patients ranged between 14 and 73 years, with 48% being over 18 years. Out of this adult group, 41% were married, and the consanguinity rate in our sample was about 38%. Of the married group, 68% had no affected children, 22% had one child with SCD, and the remaining 10% had two affected children. Approximately 73% of the SCD patients had a low level of education (intermediate or secondary school) and the majority (81%) were unskilled laborers. Interestingly, 82% of the total SCD patients thought that the disease was caused by hereditary factors, and 72% responded that the disorder was preventable if premarital counseling was available. When these patients were also asked about their opinion regarding health education, 59% considered it inadequate in Bahrain. Fifty-three percent of the patients thought that SCD was not related to any specific gender. In response to an important question about whether the blood or an organ was the cause of the disease, 64% thought that SCD was a blood disorder and surprisingly, 18% thought it was due to joint and bone disorders. When asked about the management of SCD, 53% of patients believed blood transfusion to be an effective treatment. However, 65% were not aware of the potential hazardous effects of transfusion as a source of transmitting communicable diseases such as hepatitis, AIDS, etc.

All the patients had blood tests for hemoglobinopathies which showed that 18% also had G6PD deficiency and 11% had either  $\alpha$ - or  $\beta$ -thalassemia. Hemoglobin levels ranged between 5.6-11.8 g/dL, and nearly 22% of the males and 34% of the females were severely anemic (Hb <8 g/dL). Among the 31 patients who had had blood

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transfusions, 37% were positive for hepatitis C and 11% had HBsAg.

Apart from the usual schedule of management to relieve pain, a review of the medical charts indicated that 31 patients (26%) had received blood transfusions amounting to 181 units of packed RBCs over the previous year, with males receiving more transfusions than females (76% vs. 55%). Of these patients, 27 had been transfused four or more times. During the four-month study period 23 patients were transfused, receiving 1-3 units of packed RBCs.

### Discussion

Although this study is limited in sample size, it provides important information about SCD with regards to the health knowledge of the patients. One interesting finding is that during this short four-month study period, 118 out of approximately 700-750 SCD patients in the country (approximately 17% of the total cases) were admitted to the hospital (the number of SCD patients was calculated by the Hardy Weinberg formula, using 600,000 indigenous population as base and a sickle cell gene frequency of 1.2%). This number of admissions is relatively high and should be discouraged, as many mild SCD crises can be managed in the primary health care clinics or even at home if the patients are given adequate information about their disease. Furthermore, studies have shown that in Bahrain there are many cases of either benign or clinically mild SCD. These are patients with high HbF (>10%), which has a moderating influence on the gelatin of deoxy HbS.<sup>4</sup> Forty-nine percent of our patients were found to have high HbF.

Another interesting observation was the age of many of our SCD patients. Many of them were in the higher age bracket, the oldest being 73 years of age. This is in contrast to black patients with SCD, who normally die by the second decade of life. Diggs<sup>5</sup> has reported that 25% of deaths of affected black patients occur in the first two years of life. These patients usually have a low level of HbF (<5%) and bacterial infections have been known to contribute to morbidity and mortality. In the Arabian Gulf region, two different patterns of SCD are well documented,<sup>6,7</sup> one associated with mild disease and high levels of HbF (>10%), and the other in which HbF level is low (<5%), with severe clinical manifestations. Many of our patients had unusually high levels of fetal hemoglobin (as shown in their medical charts), suggesting the presence of benign clinical disease in Bahrain, thus contributing to long life. Although there are no threshold levels of HbF which may ameliorate the clinical course of SCD,<sup>8</sup> data are available<sup>9</sup> to show that the coexistence of the  $\alpha$ -thalassemia gene with raised levels of HbF is an important predictor of clinical severity.

A cumulative analysis of the health questionnaire showed that 38% of patient had little knowledge about SCD, 32% had moderate knowledge, and the remaining 30% had a high degree of knowledge about the disease. Fifty-nine percent of the patients thought that there was not enough health education about the disease in the community. This underlines the need for a stronger health education campaign about SCD in the state of Bahrain, which will benefit not only the patient but the community at large, as it may be an important step in controlling the hereditary anemias in the country.

Regarding the clinical manifestations, 97% of our sample presented with painful crises. In most cases, these painful crises are mild and are the transient type of attack which can be taken care of at home. In this regard, the patients' education or knowledge about the disease becomes important, and should not be overlooked. In Bahrain there is no specific season in which patients are more prone to crises, but the weather from May to August is hot and humid, so patients should be advised to stay indoors as much as possible and to get sufficient rehydration, as this change of weather can lead to changes in blood viscosity, resulting in crises.

Apart from the regular regimen used for the management of their SCD, 19.5% of the patients who received blood transfusions to correct their anemia were given 1-3 units of packed RBCs. Review of the patients' charts showed that 65% of our patients had a history of repeated blood transfusions during their lifetime. The practice of routine blood transfusion should be discouraged, and should be given only when absolutely necessary. It is therefore suggested that rigid criteria should be laid down for transfusing SCD patients to minimize the wastage and hazardous effects of blood transfusion. Such criteria would be disseminated among medical practitioners by proper continuous medical education. For those patients who request blood transfusions because of false beliefs, a structured health education program should be developed to inform the public about the advantages and hazards of blood transfusion.

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