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Table 1. Causes of mortality among sickle-cell disease patients hospitalized during 1997-2005.

| Causes of mortality                     | Male | Female | Total | Percentage |
|----------------------------------------|------|--------|-------|------------|
| Acute chest syndrome                   | 13   | 9      | 22    | 28.5       |
| Salmonella septicaemia                 | 7    | 2      | 9     | 11.6       |
| Other Sepsis                           | 4    | 4      | 8     | 10.3       |
| Multiorgan failure                     | 5    | 1      | 6     | 7.7        |
| Hepatic crisis                         | 2    | 4      | 6     | 7.7        |
| Central nervous system                 | 2    | 3      | 5     | 6.4        |
| Delay hemolytic reaction               | 0    | 4      | 4     | 5.1        |
| Pulmonary hypertension                 | 2    | 1      | 3     | 3.8        |
| Malignancy                             | 1    | 2      | 3     | 3.8        |
| Chronic renal failure                  | 2    | 0      | 2     | 2.5        |
| Other medical problems                 | 1    | 1      | 2     | 2.5        |
| Unknown cause                          | 4    | 3      | 7     | 9.0        |
| Total                                  | 43   | 34     | 77    | 100%       |

Patterns of mortality in adult sickle cell disease in Al-Hasa region of Saudi Arabia

To the Editor: Sickle cell disease (SCD) is an autosomal recessive disorder characterized by production of abnormal hemoglobin S and is associated with higher morbidity and mortality because of anemia, susceptibility to infections and multiple organ dysfunctions. Lately, due to better understanding of SCD pathophysiology and better management of the complications, the life expectancy has significantly improved.1-3 The nature of SCD in the Eastern province of Saudi Arabia is benign because of high Hb F, concurrent inheritance of alphathalassemia (which may modify the severity of clinical course) and with preserved splenic functions in the majority of the patients until adulthood. This reduces the risk of severe pneumococcal infection related mortality in this region, unlike most African or American patients.4 In the Eastern province SCD is responsible for 15.7% to 21.1% of hospital admissions to pediatric and medicine wards.5

The present retrospective study was undertaken at the 502-bed King Fahad Hospital and Tertiary Care Center, Al-Hofuf, Eastern province of Saudi Arabia. Records of all the patients having SCD, admitted to the Medicine Department during the period of January 1997 to December 2005 were reviewed. All these patients were confirmed as having SCD on the basis of hemoglobin electrophoresis results, other supportive hematological parameters and past clinical history. The cause of death was established on the basis of presentation of the patient to the emergency department, the course of illness during hospitalization, supportive laboratory evidence and clinical management of these patients.

Of 10 461 admissions of SCD adult patients to the medical department during this period, 77 (0.73%) expired. Of those, 43 (55.8%) were male and 34 (44.2%) female, with a male female ratio of 1.3:1. The mean age of the male patients was 30±14 years (range, 16-67 years) and of female patients 27±13 years (range, 14-67 years). The overall mortality of 0.73% in the present study was much lower than that reported from other parts of the world.6 The majority of the deaths (51.9%) occurred among patients in the 20 to 30 years of age group and 20.7% were younger than 20 years of age. The majority of the patients had high Hb F (20% to 24%), a steady status leukocytes below 15 000/µL and no evidence...
of chronic organ damage (renal failure, stroke). The acute events leading to deaths in these subgroups were acute chest syndrome (28.5%), followed by Salmonella septicemia (11.6%) and other bacterial sepsis among 10.3% of the patients (Table 1). Acute chest syndrome as a common serious complication of SCD has been reported from this region.5-7 The Salmonella septicemia in these patients presented with a fulminating course and rapid fatality which could be either due to highly virulent nature of the Salmonella bacterium or as a result of misdiagnosed Salmonella infection as these patients presented with different manifestations in comparison to the Salmonella septicemia in patients not having SCD.8,9

Salmonella septicemia in the present study had three striking features: a preponderance among males, as 7/9 (77.7%) of the patients where Salmonella septicemia was the cause of death were male, (b) liver function abnormalities which mimicked hepatic crisis (high transaminase, high total bilirubin, mainly conjugated bilirubin), and (c) a rapid fatal course in comparison to other bacterial infections. The cause of death could not be ascertained in 7 (9.0%) of the patients. SCD patients have an increased susceptibility to bacterial infections because of defects in splenic function, opsonization, phagocytosis and sluggish activation of the alternative complement pathway. Patients with SCD have a greater predisposition for developing Salmonella septicemia perhaps because of hypersplenism and a defective complement system hindering phagocytosis and impaired macrophage function.8,10 Salmonella septicemia episodes among 42% of SCD patients in the younger age group (6-20 years), with a lower case fatality rate of 2.2% as compared to the present study, has been previously reported from the Eastern region of Saudi Arabia.9 In the present study, Salmonella septicemia episodes were more frequently (11.6%) associated with mortality of adult patients of 20 to 30 years age group.

Pulmonary hypertension, a life-threatening complication of SCD, has been reported to be a common cause of death among these patients.11,12 In the present study only three patients died because of chronic pulmonary hypertension. The incidence of pulmonary hypertension among SCD patients in this region is not known, which emphasizes the need for a screening program for early diagnosis of this serious complication. During the study period no perioperative or early postoperative mortality was observed among SCD patients undergoing surgical procedures, though exchange transfusion to lower the targeted Hb S level to 50% was started after the year 2000.

The commonest sickle cell chronic complications observed in the present study were splenic among 24 (infarction, abscess, hypersplenism), hepatic and gall bladder disease among 14 patients. Although acute chest syndrome was the commonest (28.5%) cause of death among the SCD patients in this region, Salmonella septicemia and other sepsis were the next frequent cause of mortality. There were no deaths related to malnutrition, AIDS or malaria, which reflect the role of regional socioecological factors.

Although there is a high prevalence of SCD in this region of Saudi Arabia, the overall mortality in adult patients appears to be low (0.73%) and acute events (acute chest syndrome, Salmonella septicemia and bacterial sepsis) are the major cause of deaths. The natural history of SCD and pattern of death in Saudi Arabia can be reliably assessed only through a cohort study from birth.

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