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A SURVEY OF PATIENTS WITH SICKLE CELL CRISIS PRESENTING TO ACCIDENT AND EMERGENCY DEPARTMENT OF SALMANYIA MEDICAL CENTER, BAHRAIN

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ABSTRACT

احتلت أمراض فقر الدم الوراثية في عام 1990 المركز الثالث بين عشرة أمراض استدعت مراجعة مركز السلمانية الطبي . وقد تمت دراسة وتقويم الأعراض السريرية والمراحل العلاجية ل 200 شخص بحريني مصاب بفقر الدم المنجلي ممن راجعوا قسم الطوارئ بمستشفى السلمانية خلال نوبة انسداد الأوعية الدموية.كما أن عدد الذكور فاق عدد الإناث بنسبة 1 : 2 وإن 60% من هؤلاء ضمن المجموعة العمرية (15-30) عاما وتظاهر معظم (86%) بألم الأطراف يتبعه ألم في البطن وآلام جسمية متعممة. وقد استجاب المرضى للعلاج بالاماهة ومسكنات الألم المخدر أو بالعقاقير المضادة للالتهاب غير السيترويدية وقد غادر (83%) منهم قسم الطوارئ. ويبدو أن المرضى البحرينيين مصابين بنوع متوسط من فقر الدم المنجلي.

In 1990 hereditary anaemia were found to occupy third place among ten leading causes of attendance at Salmaniya Medical center (SMC)[1]. two hundred Bahraini sickle cell disease (SCD) Patients who attended Accident and Emergency (A&E) department for vaso-occulusive crisis (VOC) during the period of January to March 1994, were studied to evaluate their clinical presentation and management. Males outnumbered females in ratio of 2:1,60%were in the age group of 15-30 years. Extermities pain was the commonest presenting feature, 86%followed by pain in abdomen and generalised body ache. Most patients responded to treatment with hydration, narcotic analgesic or non-steroid anti-inflammatory drugs and were discharged home from A&E 83% It appears that Bahraini Patients suffer from a milder type of SCD.

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Introduction

In 1990 herediatery anemias were found to occupy third place among ten leading causes of attendance at SMC. SCD are inherited disorders of haemoglobin (Hb) structure and synthesis. It includes homozygous sickle cell anaemia (Hb SS), and heterozygous. Sickle cell trait (Hb SA) and less common variants like double heterozygous combinations of HbS and B thalassemia2. sickle cell hemoglobin (HbS) is the most widely distributed mutant of normal adult haemoglobin (HBA)[3] SCD is common in many parts of the world viz. Africa, Middle East, Mediaterranean basin and South America. It drains health resources and dramatically affects family and personal life.

SCD may present as VOC, haemolytic anaemia, marrow or fat embolism, a plastic crisis, bone necrosis and priapism, heart failure, acute abdomen hebatic crisis, splenic sequestration and choleithiasis [3-7]. Severity of the presentation may vary from mild to severe requiring several blood transfusions and hospitalisations. No study has been conducted to explore the emegency presentation and management of SCD in Bahrain. Therefore we decided to conduct this study to assess the above aspects of SCD.

Subjects and Methods

Two hundred Bahrain patients of SCD attending A&E department with sickle cell crisis during the period of January to March 1994 were selected randomly by the different physicians on duty. Non-Bahraini patients were excluded from the study. A prepared questionnaire was filled by each physician. The

questionnaire included age. Sex nationality, marital status, occupations, source of referral, number of visits to A&E, clinical presentation, precipitating factors, relevant investigations like full blood count, x-rays were performed, and details of management. The management was standardised as hydration, non-steroid anti-inflammatory drugs, and in case of inadequate response narcotic analgesics. The data was summarised, tabulated and analysed.

TABLE 1
Occupation of patients with SCD and VOC

Occupation	Percentage
Student	70%
House wife	18%
Labourer	3.5%
Professional	2.6%
others	3.5%

Results

The charts of 200 Bahraini patients were studied. Figure I show that 90% patients were under 30 years of age. Male to female ratio was 2:1 figure II. The marital status showed 78% unmarried, 21% married and 1% widowed. Majority of patients were students 70%; 18% were housewives, 3.7% labourers and 2.6% professionals Table I. Self referred were 56% to A&E and the rest were referred from health centres. Nearly one third 32% of patients attended the A&E between 1:00 am and 7:00 am the rest were distributed evenly throughout the day.

TABLE 2
Clinical presentation of SCD patients Presenting to A&E.

Clinical presentation	Percentage
Pain	86%
Anaemia	38%
Jaundice	18%
Fever	20%
Hepatosplenomegaly	2%
Respiratory Disease	1.5%
Osteomyelities	1%
Other	5.5%

The main presenting symptom were pain 86% and fever 14%. Out of all patients presenting with pain, it was reported in the lower extremities in 53%, in the upper extremities 30% abdomen 11% and generalised body pain in 6% cases, Table II. 84% patients had one or two crisis per year and only 11% had more than two crisis in one year. The main precipitating factors were exposure to cold and heat in 79% and 21% cases respectively. Patients responded (83%) to treatment with hydration and non-narcotic analygestics or narcotic analgesics and were admitted to the hospital.

Discussion

In the present study we found that majority of the patients 60% were in the age group of 15-30 uears. Bonadio in saudi Arabia also found higher incidence of painful crisis in patients between the age of 2.5-18 years[7]. A study conducted by lester reported higher incidence of painful crisis and complication between the age of two months to 18 years[6]. This is because this is the

active period of life with increased physical activity. During hot and humid weather they loose water and salt in sweat and are prone to vaso-occlusives crisis, 77% of the patients were un-married and 78% were students. Besides being in the age groub of studentship, if they have emotional and psychological factors operating, needs to be evaluated in further studies.

In this study we found that male to female ratio of patients was 2:1 Mulik et al in their study of 99 patients and had 60 males and 39 females[9]. Bonadio in study of 43 patients reported 25 male and 18 females[7], whereas Lester et al in a study of 64 children reported 35 girls and 29 boys, thus males are predominant suffers from VOC in most of the studies.

It probably shows that females are having milder course of the disease then males., Does the Heamoglobin in two sexes behave differently; or coexistence of hertocellular hereditary persistence of Hbf may ameliorate the vaso-occulsive crisis, or presence of X-liked gene at position XP 22-2 plays some role to protects females remains to be evaluated further[8].

Pain was the main presenting symptoms in these cases 86% and fever in 14%. Pain in the upper and lower extremities constituted 83% of the cases. Bonadio reported 42 episodes 45% of pain in the extremities in 106 episodes of painful crisis[7]. Mulik et al reported hand and feet syndrome in 58% of the sickle cell crisis in western province of Saudi Arabia[9]. El-Hazmi from South Western province of Saudi Arabia reported hand feet syndrome in 34% of the cases[10]. Walson reported 11% of hand and feet syndrome in Black Americans[11], 32% of the patients attend the hospital between 1 am to 7 am and the rest were equally distributed over the period of 24 hours. Usually hot and humid weather is known to cause the crisis but cold blow of air conditioner and cold weather may also precipitate the crisis. Mulik reported increase in crisis and hospital admissions in month of December[9]. non-availability of health center services during this period, may be the other

reason for increased attendance between 1-7 am. 83% of the patients responded to hydration by intravenous fluids, analgesics or narcotic therapy and only 17% needed admission. The main reason for admission was vaso-occulsive crisis. It appears that the disease runs a milder course in Bahraini population than in black Americans. Patients from Western Coast of Saudi Arabia have been reported to have milder course too; where one in five patients are hospitalised[10], their figures are similar to or data and sows influence of the geographical distribution of the disease and similarity of clinical presentation in two neighboring countries. There were on an average two crisis per year, 11% patients had more than two crisis per year, 25% of the patients had on an average one blood transfusion.

Conclusion

Since no specific and definitive therapy for sickle cell disease exist currently, oral of parental hydration and narcotics remains the main stay of therapy of acute painful crisis refractory to conservative care. Sickle cell crisis can be managed successfully in the health centers and only refractory cases should be sent to accident and emergency department. More studies are needed on the socio-economic factors and behaviour of hemoglobin's in male and female with special attention to X linked influence.

REFERENCES

1. Bahrain Health information Center, health statistics abstract, Government press, Ministry of Health, Bahrain. 1990;115
2. Froncis RB, et la. Vascular occlusion in sickle diseases. Current concept and unanswered questions blood 199;777:1405-1414.
3. El-Hazmi MA, et al. Features of sickle cell disease in Saudi Children J. Trop. Pediatr 1990;364:479-481.

4. Godeau B, et al. A plastic crisis due to extensive bone marrow necrosis and previous infections in sickle cell disease. Am. J. Med 1991;91 5:557-558.
5. Hamre MR et al. Priapism as a complication of sickle cell disease (case report) J. Urol. 1991; 145 1:6-68.
6. Lester L.A. et al. Cardiac abnormalities in children with sickle cell anaemia. Chest 1990;98 5:1169-1174.
7. Bonadio WA. Clinical features of abdominal painful crisis in sickle cell anaemia. J. pediar surg. 1990;253:301-302.
8. Rodgers GP. Recent approaches to the treatment of sickle cell anaemia (clinical conference), JAMA 1991;265 16:2097-101.
9. Mulik R.Et al. Clinico-epidemiological study of sickle cell anaemia in Saudi Arabia. J. Trop Pediatric c 1991;37 3:100-105.
10. El-Hazmi MAF and Sarys AS. On the nature of sickle cell disease in the south western province of Saudi Arabian acta Haematal 1986;76:212-216.
11. Watson RF, et al. The hand foot syndrome in sickle cell disease in young children. Pediatric 1963;31:975-982.