

Review

Adult sickle cell diseased patients' knowledge and attitude toward the preventive measures of sickle cell disease crisis

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Sickle cell crisis is preventable most of the times. However, in Bahrain sickling crisis is the most complained and cause of hospital admission. No data is available regarding the extent of knowledge and attitudes towards the sickling crisis's preventive measures the adult Bahraini sickle cell patients have. The purpose of this paper is to explore the extent of knowledge and attitude of Bahraini adult sickle cell patients towards the preventive measures of sickle cell crisis. The objective is to establish baseline data and to utilize the findings in designing awareness programs that would assist the sickle cell patients to prevent the sickling crisis. A convenient sample of 84 Bahraini adult sickle cell patients achieved. Structured interview approach was used to assess the subjects' knowledge and attitudes toward the preventives measures of sickle cell crisis. The results indicated that the sample was moderately knowledgeable about the sickling preventive measures (Mean of Knowledge score = 55%) and moderately compliant (Mean of Attitudes score = 63%). The subjects' knowledge (about the preventive measures) was found to be moderately and positively correlated ($r = 0.57$, $r^2 = 0.32$, $p = 0.000$) with their attitudes toward crisis prevention. Designing awareness programs regarding the preventive measures of sickle cell crisis is an evident need for these patients.

Key words: Sickle cell disease (SCD), sickle cell trait (SCT), sickle cell disease crisis, sickling crisis, Vaso-occlusion crisis (VOC), knowledge, attitude, preventive measures of crisis.

INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive disorder in which abnormal hemoglobin leads to chronic hemolytic anemia with numerous clinical consequences (Papadakis et al., 2006). It considered as one of the most prevalent hereditary hematological disease worldwide (Lourerio and Rozenfeld, 2005).

Worldwide, about 300 000 children are born with sickle cell disease every year (Okpala et al., 2002). It is found in many parts of the world, particularly in people whose ancestors come from sub-saharan Africa, India, Saudi

Arabia and Mediterranean countries (Turkey, Italy, Greece) (World Health Organization, 2006).

According to Bahrain Health Statistic 2004 - 2005, the first reason of admission to Salmaniya Medical Complex SMC was found to be cause by SCD. The number of hospital admission due to SCD gradually increased with 2600 patients admitted during 2005, which accounted for 6.5% of the total admission to SMC.

A key issue in managing sickle cell disease patients is the early identification of high risk subjects for poor outcome, in order to initiate treatment prior to the development of debilitating organ damage (Schnog et al., 2004). The objective of this article is to evaluate the extent of SCD patient's knowledge and attitude toward prevention of sickle cell crisis that may help concerned health professionals in designing educational programs which emphasize in dissemination of sickling crisis preventative measure

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Abbreviations: SMC, Salmaniya Medical Complex; SCD, Sickle cell disease; SCT, Sickle cell trait; VOC, Vaso-occlusion crisis.

Ethical consideration

The research committee (SMC, Ministry of Health) and the nursing administration of SMC approved the study and they send the approval letter to the concerning wards. The researchers maintained the anonymity and confidentiality of the participants after informed consent obtained.

Problem statement

What is the extent of adult sickle cell diseased patients' knowledge and attitude toward the preventive measures of sickle cell disease crisis?

Purpose

To evaluate the extent of sickle cell diseased patients' knowledge and attitude toward prevention of sickle cell crisis.

Objectives

1. To help the concerned health care professionals in designing educational programs which aim to increase the sickle cell patients' awareness and enhance their attitude toward the preventive measures of sickle cell disease crisis.
2. To assist sickle cell patients (through community based organizations) to participate in awareness programs concerning crisis prevention.
3. To suggest recommendations that focus on the involvement of public and health sectors (health centers, schools, associations, etc...) those will emphasize on dissemination of sickling crisis preventive measures.

Benefit of study

To overcome the deficiencies of patient's knowledge and attitude regarding preventive measures of SCD crisis, which may help to reduce morbidity and mortality rate. Ultimately, achieving these goals will help to reduce the total cost of Ministry of Health (MOH) budget.

Importance of the study

From our work experience in taking care of SCD patients in adult intensive care unit (ICU), we observed that these patients are at high risk of developing severe complications of sickle disease crisis, which are for example: Cerebrovascular accident (CVA), acute chest syndrome, septic shock and eventually multi-organ failure. Obviously,

the SCD crisis consequences might be preventable if the SCD patients become adherent to the preventive measures of SCD crisis.

In 2005, Salmaniya Medical Complex (SMC) recorded that adult patients (≥ 15 years) with principal diagnosis of sickle cell disease have the highest number of discharge ($n = 2555$), days of care ($n = 9255$) and average length of stay ($x = 4.6$ days) compared to any other disorders.

In year 2007, the average number of sickle cell patients admitted with crisis was 232 monthly. These patients required 1153 days of hospital care with 5 days length of stay on average each month (Admission Office SMC, 2007).

Literature review

No study was found investigating the knowledge and attitude of sickle cell patients toward the preventive measures of sickling crisis. However, few studies investigated the sickle cell patients' general knowledge about their disease.

Research Reports Al-Nasir and Niazi (1997), regarding the general knowledge of 118 SCD patients and found: 38% had little knowledge about SCD, 32% had moderate knowledge and 30% had high degree of knowledge.

The author also recommended that: first, mild SCD crisis can be managed in the primary health care clinic or at home if the patient is given adequate education about their disease. Second, there is need for a strong health education campaign about SCD in Bahrain.

A study done among African American students about awareness of SCD by Ogamdi (1994), found that the subjects were not aware of the basic facts about sickle cell disease.

As mentioned in a study done by Al-Arrayed (1994), on school age students' about factors participating in SCD crisis founds that cold is 45%, fever is 35%, exhaustion is 35%, temp change is 19%, hot weather is 10%, crowd is 10%, psychological tension is 10%, vomiting and diarrhea is 7% and traveling by air is 1%.

According to literature, 11 areas were identified as domains that contribute to preventive measures of SCD crisis.

These domains are general precaution, exercise, food, rest, travel, places, weather, medication, emotional, social, spiritual and coexisting medical condition. It's mean if participants have any other hereditary diseases that may aggravate sickle cell crisis (e.g. thalasemia, G6PD reduced activity or other condition like pregnancy).

General precautions

This domain encompasses several areas sickle cell patient might undertake to limit the crisis occurrence. Vaccinations during childhood against *Streptococcus*

pneumonia, *Haemophila influenzae* and penicillin prophylaxis have dramatically reduced infection related mortality in SCD patients (Schnog, 2004). Improved preventive measures such as pneumococcal vaccines for infants may reduce the number and cost of sickle cell related hospitalizations in the future (Onley, 1999).

Frequent and comprehensive medical examination should be guided by ongoing consultation with hematologists specializing in sickle cell disease. Eye examination from age of 10 years is necessary as well (National Institute of Health, 2002). Cigarette smoke, both active and passive smoking can damage the lungs and lower oxygen levels in the blood for children and adults who have sickle cell disease (WebMd, 2005). In addition, a link between cigarette smoking and "acute chest syndrome" in sickle cell anemia is suggested (Young, 1992).

Rest and activity

Keeping balance between activity and rest is crucial for sickle cell patients. Low impact exercise like leg lift and light weights may be useful and safe for maintaining strength particularly in the legs and hip but patients should consult their doctor about any exercises program. (WebMd, 2005).

In a randomized controlled trial conducted by Meremikwu (2006), it was found that moderate exercise is generally accepted to be beneficial, especially in reducing the risk of cardiovascular disease. Moderate exercise is therefore unlikely to cause harm in people with sickle cell disease. Strenuous exercise is suspected to lead to factors that may precipitate sickle cell crisis, such as low tissue oxygen saturation, dehydration and stress. People who have this disease should try to avoid activities that reduce the amount of oxygen in their blood.

Diet

Consuming nutritious food is helpful in sickling crisis prevention. In a study viewed SCD opinion regarding triggering factors of sickle cell crisis, almost 55% of the subject mentioned beans including broad beans as the participating cause. An improvement in the patient's condition was noted with increased fluids, fruits, vegetables and milk consumption (Al-Arrayed, 1994).

Places and traveling

Presenting in certain places would trigger the sickling crisis as well. Flying in an unpressurized airplane can cause cells to sickle (WebMd, 2005). Anything that reduces the amount of oxygen in their blood, such as mountain climbing, flying at high altitudes without sufficient oxygen may bring on a sickle cell crisis.

Weather

The impact of weather on sickle cell patient is obvious. Hot and humid weather make SCD patients lose water and salt in sweat and they are prone to vaso-occlusive crisis (Al- Arrayed et al., 2007).

Medication

Only hydroxyurea has been proven to reduce the incidence of painful crisis and it safe and effective in preventing complications (Schnog, 2004). World Health Organization (2006) added that treatment with hydroxyurea has reduced many of the major complications.

Psychological status

Psychological and emotional status can largely influence sickle cell patient health. Painful crisis are preceded by increases in severe stressors 2 days prior to onset. Patients with SCD have reported social problems including employment, finance, childcare and spare-time activities. Results showed that patients who were able to maintain positive affect were less likely to use health care services despite experiencing pain (Porter et al., 2000).

A report from Mayo clinic USA in (2007), emphasized that stress can be reduced and avoided by praying, seeking help from family and friends. Having a positive attitude, creating a supportive environment and develop coping skills to help to deal with the disease. Strong family relationships and close personal friends can be helpful. A support group might help to cope with the disease (American Academy of Family Physicians, 2002).

Coexisting conditions

Painful crisis occur with increased frequency in pregnant women (Schong, 2004). In pregnant women, the crises were precipitated by urinary tract infection, respiratory tract infection, sore throat and septicemia (Al Mulhim, 2000).

Theoretical framework

Operational definition of:

Knowledge

The information that SCD patient have about the preventive measures of SCD crisis, it is measured in percent from 0 - 100%, 0 means patient don't have any knowledge regarding preventive measures of SCD crisis, 100% means that patient have all information required to prevent SCD crisis.

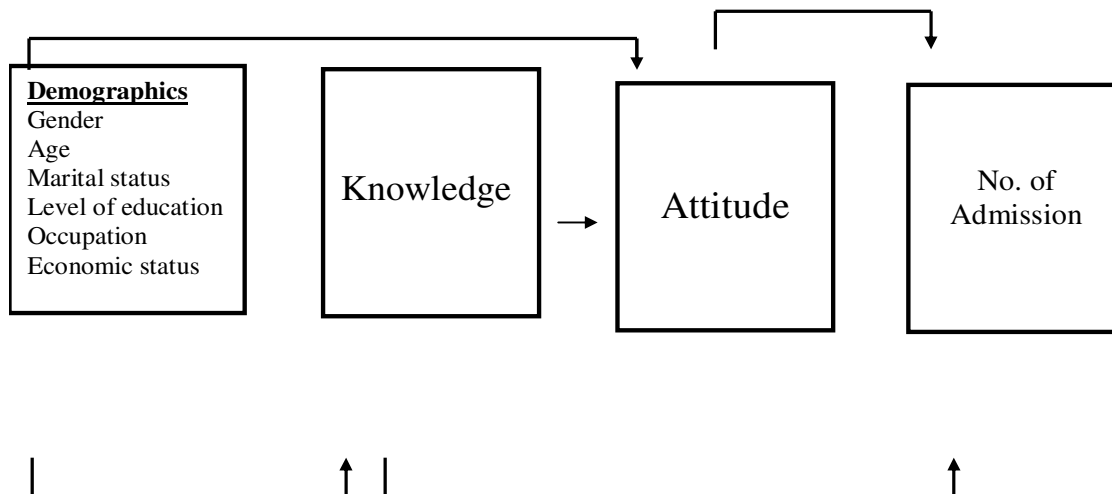


Figure 1. Theoretical framework.

Attitude

The actions those carried out by SCD patient to prevent SCD crisis, it is measured in percent from 0 - 100%, 0 means patient don't perform any action that will help in preventing SCD crisis, 100% means that patient perform all actions required to prevent SCD crisis.

Number of admission

Frequency of SCD patient's admission to the SMC hospital per year due to SCD crisis (Figure 1).

Research questions

1. What is the extent of knowledge among adult sickle cell diseased patients about sickling crisis preventative measures?
2. What is the attitude of sickle cell diseased patients toward sickling crisis preventive measures?
3. Is there a relationship between patients' Knowledge regarding sickling crisis preventive measures and their demographic characteristics?
4. Is there relationship between patient attitude toward sickling crisis preventive measures and demographic characteristics?
5. Is there relationship between patient knowledge and attitude toward preventive measures?
6. Is there a relationship between the patient's attitude toward SCD crisis prevention and frequency of admission due to SCD crisis?

Study design

1. Design type: quantitative, descriptive and correlational

study.

2. Population: convenient sample of approximately 100 subjects within the hospital, however, power analysis employed to determine the actual required sample size.

3. Subjects' inclusion criteria: see the Table 1.

4. Instrument: questionnaire was used to evaluate the subjects' knowledge and attitude toward prevention of sickling crisis, the data collected through structured interview. This tool is based on literature review of established questionnaires; however, the research team has developed the vast part of it. The validity of the questionnaire content was verified by experts opinions in the related field (hematology, intensive care) and final version achieved. The reliability measured using Chronach Alpha after data collection done.

5. Data analysis: the scores of knowledge and attitude domains calculated as the number of correct response divided by total number of each domain questions then this ratio expressed in percentage. After the scores of knowledge and attitude are obtained correlational procedures used to explore the relationship between knowledge, attitude, demographic characteristics and frequency of admission due to crisis.

Data analysis

Instrument validity

The instrument validity (construct and content) was established by specialists in the relevant field for the knowledge and attitude scales.

Instrument reliability

A pilot study performed recruiting 33 subjects revealed

Table 1. Subject inclusion criteria.

Disease status:	Diseased
Age	≥ 15 years
Gender	Any
Education level	Any
Occupation	Any
Level of activity	Any
History of crises	Any
Family history	Any
Setting	In-hospital

Table 2. Details of subjects' demographic characteristics.

Demographic variable		n	%	
Gender	Male	40	49	
	Female	42	51	
Marital status	Single	50	59.5	
	Married	33	39.3	
	Widower/Widow	1	1.2	
Level of education	Illiterate	3	3.6	
	Up to high school	52	62.7	
	University	28	33.7	
Occupation	Student	27	32.5	
	Unemployed	26	31.3	
	Employed	29	34.9	
	Retired	1	1.2	
Economic status	Less than 200 BD	13	15.5	
	200 - 500 BD	61	72.6	
	More than 500 BD	10	11.9	
Age				
Mean	St. Dev.	Minimum	Maximum	Range
27	9.2	15	59	44

reliable knowledge scale as Cronbach Alpha = 0.74 and Attitude scale with Cronbach Alpha = 0.72

RESULTS

Demography

The sample consisted of 84 Bahraini adult sickle cell diseased patients of which male (49%) and female (51%). The majority of the sample was single (60%), up to high school education (63%) and earns monthly between 200 to 500 BD (73%). Table 2 lists the demographic characteristics in details:

Research question 1 result

The extent of knowledge was estimated as the ratio be-

tween the correctly answered questions and the total asked questions. The sample was found to be moderately knowledgeable about the preventive measures of sickling crisis as their mean knowledge score was (55.3%) (Table 3).

This mean knowledge score was explained by the high percentage of subjects who incorrectly answered nearly half of knowledge scale questions (Table 4).

Cluster analysis employed to reveal any natural groups that might exist based on the subjects Knowledge Score. K-Mean cluster analysis was able to segregate 3 clusters (groups) who were significantly different in their means of Knowledge Score. One-way Analysis of Variance (ANOVA) was used to confirm the significant difference between groups' means. The analysis showed that each group mean was significantly different from each other as ($F(2, 81) = 120.5, p = 0.000$). Group membership exerts large effect size on the Knowledge Score as

Table 3. Descriptive statistics for subjects' knowledge score.

Statistics on sample Knowledge Score					
Mean	St. Dev.	Minimum	Maximum	Range	CI 99%
55.3	12	20	85	65	51.8 - 58.7

Table 4. The nine questions to which most subjects incorrectly answered.

Question	Domain	n	%
1. Which of the following vaccine will help to prevent the sickling crisis?	General precautions	78	92.9
2. It is important to have nutritious meals rich in protein because:	Food	78	92.9
3. Which of the following precautions might be helpful to prevent the long-term complications of SCD?	General precautions	69	82.1
4. While traveling in airplane the following factors might contribute to SCD crisis:	Travel	68	81
5. Which of the following food is rich source of iron?	Food	60	71.4
6. Which of the following will trigger SCD crisis?	General precautions	58	69
7. Which of the following places should be avoided by SCD patients:	Places	58	69
8. It is advisable for SCD patient to attend all the following social activities, except:	Social activities	43	51.2
9. It is not advisable for SCD patient to be in highly crowded places because:	Places	42	50

Table 5. Statistics on Knowledge score for each group.

Cluster	Mean	St.Dev.	Minimum	Maximum	Range	CI 95%
Poorly Knowledgeable	35.5	8.5	20	43.8	23.8	29.8 - 41.3
Moderately Knowledgeable	53.8	5.5	45	61	16	52.3 - 55.3
Highly Knowledgeable	70.8	5.9	63	85	21	68 - 73.6

Score as Eta Squared was = 0.74.

The first group size was 11 cases (13% of sample) with mean Knowledge Score of 35.5%. The only demographic characteristic that made this group unique was that up to high school education subjects were significantly more than university educated counterparts (chi = 6.16, DF = 1, p = 0.013). We would label this group as "Poorly Knowledgeable Group".

The second group was the majority of the subjects. The size was 54 cases (64.2%) with mean Knowledge score of 53.8%. No unique demographic characteristic was isolated for this group. This group labeled as "Moderately Knowledgeable Group".

The third group was 19 cases making 22.6% of the

entire sample. Their mean Knowledge Score is 70.8%. Like the second group, no unique demographic characteristic was isolated. This group labeled as "Highly Knowledgeable Group" (Table 5).

Research question 2 result

The extent of compliance was estimated as the ratio between the reported compliant attitude and the total asked questions. The sample was found to be moderately compliant with the preventive measures of sickling crisis as their mean attitude score was (64%) (Table 6). Cluster analysis employed to reveal any natural groups

Table 6. Statistics on the Attitude score.

Statistics on sample Attitude Score					
Mean	St. Dev.	Minimum	Maximum	Range	CI 99%
64	13	27	94.2	67.2	60.2 - 67.7

Table 7. Statistics on Attitude score for each group.

Cluster	Mean	St. Dev.	Minimum	Maximum	Range	CI 95%
Poorly Compliant	43	8.3	27	51	24	38 - 48
Moderately Compliant	62	5	53	70	17	60.5 - 63.5
Highly Compliant	79.2	5.5	70	94	24	76.8 - 81.5

that might exist based on the subjects Attitude Score. K-Means cluster analysis was able to segregate 3 clusters (groups) who were significantly different in their means of Attitude Score. One-way Analysis of Variance (ANOVA) was used to confirm the significant difference between groups' means. The analysis showed that each group mean was significantly different from each other as ($F(2, 81) = 108, p = 0.000$). Group membership exerted large effect size on the Attitude Score as Eta Squared was = 0.72.

The first group size was 13 cases (15.4% of sample) with mean Attitude Score of 43%. No unique demographic characteristic was isolated for this group. We would label this group as "Poorly Compliant Group".

The second group was the majority of the subjects. The size was 47 cases (56%) with mean Attitude score of 62%. 2 unique demographic characteristics were isolated for this group. First, singles are significantly more than married ($\chi^2 = 4.35, DF = 1, p = 0.036$). Second, up to high school educated are significantly more than university educated ($\chi^2 = 5.10, DF = 1, p = 0.023$). This group labeled as "Moderately Compliant Group".

The third group was 24 cases making 28.5% of the entire sample. Their Attitude Score was 79.2%. Like the first group, no unique demographic characteristic was isolated for this group. This group labeled as "Highly Compliant Group" (Table 7).

Research question 3 result

The knowledge and the attitude score were found to be strongly correlated ($r = 0.57, p = 0.000$) in non-linear fashion. A cubic curve was best describing the relationship between them. The Attitude Score was predictable using the Knowledge Score using the following cubic equation:

$$\text{AttScr} = -90 + (10 \times \text{KnowScr}) + (-0.196 \times \text{KnowScr}^2) + (0.001 \times \text{KnowScr}^3)$$

Research question 4 result

Multifactor between-groups ANOVA performed to explore the impact of age, gender, marital status, education level, occupation and economic status on the subjects Knowledge Score. The analysis revealed non significant model ($F(34, 4121) = 0.945, p = 0.545$). None of the demographic variables showed significant Main effect or Interaction effect over the Knowledge score. Except for a subgroup in the education level, the university educated subjects (mean \pm SD, 61.2 ± 11.8) was significantly more knowledgeable about the preventive measures compared to up to high school educated counterparts (mean \pm SD, $52.7 \pm 10.9, t_{78} = -3.2, p = 0.002$). The magnitude of the difference between the means was small (Eta squared = 0.012).

Research question 5 result

Multifactor between-groups ANOVA performed to explore the impact of age, gender, marital status, education level, occupation and economic status on the subjects Attitude Score. The analysis revealed non significant model ($F(34, 5487) = 0.87, p = 0.653$). None of the demographic variables showed significant Main effect or Interaction effect over the Attitude score.

Research question 6 result

Overall, no relationship was found between the Attitude score and frequency of crisis occurrence. However, a strong relationship ($r = -0.88, p = 0.000$) uncovered in a subgroup who had 20 to 35 crises during the last 3 years. The group size is small ($n = 13$). These results are powerful 90% as beta = 10% at alpha = 5%.

DISCUSSION

Our results came consistent with the studies that said

SCD patients mostly lack knowledge about their disease (Mean score (general knowledge) = 58%, Median = 63%). Al-Nasir and Niazi (1997), found that 70% of SCD patients will be having little or moderate knowledge about their disease. Adding to that Ogamdi (1994), affirms that "The subjects did not well understand the basic facts about sickle cell disease". Moreover, Butler (1993), found also that "in general, group members evidenced a poor understanding of sickle cell disease assessed by pretest". Integrating these findings, we found the need for educating SCD patients about their illness is evident.

A study done by Gil (1989), stated that "the acquisition of knowledge about a disease is important in behavior adaptation, especially if it is paired with believe that ones behavior will have positive impact on health". This statement explains the importance of knowledge to affect attitude (behaviors and believes). Affirming our findings that subject's knowledge regarding SCD preventive measures is positively correlated with this attitude toward it. The correlation between them was moderately strong ($r = 0.57\%$, $p = 0.000$).

SCD patients apparently share similar believes and misconceptions. About 99% of our sample believes that avoiding broad bean is a preventive measure to SCD crisis. Al Arrayad (1994), found also that 55% of the SCD patients believe that broad bean can aggravate the SCD crisis. However, no adequate evidence found regarding the relationship between broad bean consumption and occurrence of sickling crisis.

Recommendation

The SCD crisis consequences will be preventable if the SCD patients become adherent to the preventive measures of SCD crisis.

Based on the research results we suggest implications for patient to take the responsibility of self-education about SCD crisis prevention involve in social activities designed for educating SCD patients, be an active member in SCD association, utilize all available resources effectively to reduce the incidence of SCD crisis.

Implications for practice

1. Create unique Sickle Cell Crisis management protocol to be followed by health care services caring for SCD patient with holistic management.
2. Specialized Nurses to care SCD patients.
3. Provide counselor for the in-patients.
4. Provide educational program for the nurses caring of SCD patients.
5. Provision of educational materials on: (vaccination, nutrition, medication, disease complications).
6. Patients and their families should received information about the crisis as a part of ongoing treatment.

7. Provide special unit capable to manage patients with sickle cell disease crisis
8. Regular check-up

Implications for society

1. Involve the social/religious places to educate the community about the SCD.
2. Involve social/religious people in the educational programs regularly.
3. Donate special consideration for SCD patients as students and employees in terms of: type of job, level of activity, etc.

Limitation

The study sample size was small so it is recommended to have a bigger sample to study the relationship between demographic characteristics and Knowledge and Attitude. Also, the frequency of admission was obtained from the patients themselves as it was difficult to obtain it from the medical record. In addition, the data collection was depending on the patient health condition as it was difficult to interview some patients who were having pain especially in the first 2 days from admission.

Conclusion

To conclude, this study illustrated the extent of knowledge and attitudes Bahraini adult sickle cell patients have regarding the preventive measures of sickling crisis. The link between their knowledge and attitudes was uncovered suggesting the importance of educating these patients about crisis prevention.

Designing awareness programs regarding the preventive measures of sickle cell crisis is an evident need for these patients. Increasing the awareness of sickle cell patients regarding this matter will potentially affect their attitudes toward crisis prevention, thus, the chance of suffering the crisis.

REFERENCES

- Admission Office (2007). Principle diagnosis report (ICD10): sickle cell disease. Salmaniya Medical Complex. Kingdom of Bahrain
- Al – Arrayed SS (1994). The nature of sickle cell disease in Bahrain. *J. Bahrain Med. Soc.* 6(3): 125-130
- Al- Arayyed, S, Hamza A, Sultan B (2007). Neonatal Screening for Genetic Blood Disease. *Bahrain Med. Bull.* 29(3): 88- 90
- Al Mulhim K (2000). Pregnancy in sickle cell disease in the Al Hassa region of Saudi Arabia. *Ann. Saudi Med.* 20(5-6): 471-476
- Al – Nasir F, Niazi G (1997). Sickle cell disease: patients' Awareness and management. *J. Saudi Med.* 18(1): 63 – 65
- American Academy of pediatrics (2002). Health supervision for children with sickle cell disease. *Pediatric.* 109:526-535
- Bahrain Health statistics 2004-2005. Ministry of Health, Kingdom of Bahrain. p. 25

- Butler, Dennis, J, Beltran, Lou, R. (1993) Functions of an adult sickle cell group: Education, task orientation, and support. *Health and Social Work*, 18(1): 49 – 56
- Gil, KM, Abrams MR, Phillips G, Keefe FJ (1989). Sickle Cell disease pain: Relation of coping strategies to adjustment. *J. Consult. Clin. Psychol.* 57: 725 – 731
- Loureiro MM, Rozenfeld S (2005). Epidemiology of sickle cell disease hospital admissions in Brazil. *Rev Saude Publica*, 39(6): 1- 6
- Mayo Clinic Staff (2007). Sickle cell anemia. Mayo Clinic. www.mayoclinic.com/health/sickle-cell-anemia/ds00324
- Meremikwu M (2006). Sickle cell disease. *Clin. Evid.* (15):45-59
- National institutes of health. National heart, lung, and blood institute. The management of sickle cell disease. 4th edition, Bethesda, Md: The institute, 2002; NIH publication no. 02-2117
- Olney RS (1999). Preventing Morbidity and Mortality From Sickle Cell Disease. A Public Health Perspective. *Am. J. Prev. Med.* 16(2): 116 – 121
- Ogamdi S (1994). African American students' awareness of sickle cell disease. *J. Am. Coll. Health* 42(5): 234-236.
- Okpala, I., Thomas, V., Westerdale, N., Jegede, T., Raj, K., Daley, S., Costello-Binger, H., Mullen, J., Rochester- Peart, C., Helps, S., Tulloch, E., Akpala, M., Dick, M., Bewley, S., Davies, M. and Abbs, I. (2002). The comprehensive care of sickle cell disease. *Eur J. Hematol.* 68: 157 – 162
- Porter LS, Gil KM, Carson JW, Anthony KK, Ready J (2000). The role of stress and mood in sickle cell disease pain. *J. Health Psychol.* 5(1): 53-63.
- Schong, J. B., Duits, A., J., Muskiet, F., A., J., Cate, H., Rojer, R., A. and Brandjes, D., P., M. (2004) Sickle Cell Disease: A general overview. *J. Med.* 62(10): 364 – 374
- Tierney LM, McPhee SJ, Papadakis MA (2006). *Current Medical Diagnosis & Treatment.* (45th Edition), Lange. U.S.A.
- World health organization (2006). Sickle cell anemia. Fifty – Ninth World Health Assembly: Provisional agenda item 11.4
- WebMD (2005). Sickle cell disease: what increases your risk. WWW.webmd.com. Healthwise incorporate.
- Young RC, Rachal RE, Hackney RL, Uy CG, Scott RB (1992). Smoking is a factor in causing acute chest syndrome in sickle cell anemia. *J. Natl. Med. Assoc.* 84(3): 267-271