Study of awareness about complications of sickle cell disease during pregnancy in Jeddah City

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Abstract

Background: Pregnancy in Sickle Cell Disease (SCD) is at very high risk. Major steps for prevention are to carry out various programs, surveys, educating and increasing awareness about the disease and its consequences including morbidity and mortality. So, the aim of the current study was to assess the awareness about complications of SCD during pregnancy in Jeddah city, Saudi Arabia.

Method: An observational cross-sectional study was carried out on 410 participants from the general population in Jeddah city, Saudi Arabia in April 2019. The tools used in this study consisted of self-administered questionnaire divided into three sections (socio demographics, perception of SCD, and Knowledge about SCD and its complications).

Results: The study sample included 410 participants, most of them female (91%) and 9% male. More than 50% of participants were aged between 20 to 30 years old and about 59.9% were married. 73.4% were university students. Regarding detection of the level of knowledge which was measured through the scoring applied the study found that most of the participants had moderate knowledge about SCD (>60%) and there was no difference regarding socio-demographic characteristics.

Conclusion: Overall knowledge about SCD and its complications during pregnancy was moderate. So, we recommend health education programs about all aspects of SCD which should be designed, implemented and evaluated among general populations.

Key words: Knowledge, Complications, Sickle Cell Disease, Pregnancy.

Introduction

Sickle cell disease (SCD) is a hemolytic anemia characterized by abnormally shaped (sickle) red blood cells (RBCs), which are removed from the circulation and destroyed at increased rates leading to anemia (1).

Abnormality in RBCs shape is mainly attributed to the presence of hemoglobin S, which, when deoxygenated, becomes relatively insoluble and forms aggregates with other hemoglobin molecules within the RBCs (2).

A point mutation in the gene coding the β chain of the hemoglobin molecule results in a single amino acid substitution (valine for glutamic acid), which leads to hemoglobin S (3).

SCD is one of the most common genetically inherited diseases affecting mainly African Americans. In addition, it is a prevalent disorder among those from the Mediterranean area like in Turkey, and the Arabian Peninsula (4).

This disorder affects millions of individuals worldwide and is prevalent in an estimated 90,000 individuals in the United States, primarily individuals of African American descent, as well as those from regions of South America, the Caribbean, and Central America and individuals of Middle Eastern ancestry or from the Mediterranean region (5).

SCD occurs in one of every 500 African American births, and an estimated one of every 12 (2 million African Americans) have the sickle cell trait (6).

Recently, Saudi Arabia has been reported to have an increasing prevalence of SCD. The carrier status for SCD ranged from 2% to 27%, and up to 1.4% had SCD in Saudi Arabia (7).

In addition, the prevalence of consanguinity ranges from about 60% in Saudi Arabia up to 90% in some Bedouin communities (8).

The pathologic hallmarks of the disease are vaso occlusion, chronic hemolysis and increased erythrocyte adhesiveness to vascular endothelium (9).

SCD is believed to be a tetrad of pain syndromes, anemia and its sequelae , organ failure (including infection), and comorbid conditions, with pain dominating the clinical picture, and it may either be spontaneous or be triggered by the other three components of the tetrad (10).

The patient suffering from sickle cell anemia develops blood related complications and can be suspected due to a family history or by conducting clinical examination. But confirmation of a case can only be carried out by laboratory investigation (11).

Complications of SCD include serious infections, damage to vital organs, stroke, kidney damage, respiratory problems, bone marrow failure, growth failure, and cognitive impairment, maturational delay in children as well as high maternal and fetal morbidity and mortality. Recurrent complications interfere with the patient's life, especially with regard to education, work, and economic, social and psychosocial development (12).

SCD carriers are absolutely normal and healthy like any healthy person and do not know that they are carriers unless they have a special blood test, HbS electrophoresis (13).

Pregnancy in SCD is at very high risk. Many reports have documented a considerable maternal risk of morbidity and mortality and high perinatal adverse outcomes (14).

Recently, Oteng-Ntim et al., in a systematic review and meta-analysis of previous observational studies, have quantified this risk. They showed that women with SCD have an increased risk of preeclampsia and maternal death, stillbirths, preterm deliveries, and small for- gestational-age newborns (15).

Knowledge of these risks has contributed to the implementation of a multidisciplinary management program including the early detection and treatment of complications during pregnancy and postpartum, follow-up by an obstetric team and a sickle cell team, appropriate pain management protocols, and transfusion programs adapted to each pregnant patient (16).

Regarding SCD treatment, routine general prophylactic and corrective measures have been associated with marked improvement in life expectancy and quality of life among sickle cell disease patients in developed nations, which points to the importance of providing proper information on SCD. Early community-based surveys conducted on African Americans in large urban areas demonstrated limited awareness of SCD in these communities (17).

Major steps for prevention is to carry out various programs, surveys, educating and increasing awareness among the people so that maximum active participation from the population can be achieved, which is vital. Success cannot be achieved without people actively involved and showing willingness towards limiting the disease and its consequences including morbidity and mortality which can improve quality of life among the population (11).

Aim of the work: Assessment of the awareness about complications of sickle cell disease during pregnancy in Jeddah city, Saudi Arabia.

Objectives: To evaluate knowledge of complications of sickle cell anemia during pregnancy among the general population in Jeddah city.

Rationale: There is a lack of information among the general population about sickle cell anemia and its complications especially during pregnancy. Therefore, community awareness about SCD and its complications during pregnancy can enhance the control of the disease and prevent its adverse outcomes.

Patients and methods

This was an observational cross-sectional study done among the general population aimed to study the awareness about sickle cell anemia and it's complication especially during pregnancy.

A-Technical design

1. Study design:

An observational cross-sectional study

2. Study setting:

Jeddah City, data collection took place in April 2019.

3. Target populations:

Included general population as self-administered questionnaire was applied in a Google form which had been loaded on the internet.

♦ **Inclusion criteria:**

- All ethnic groups
- Any age group (20 to more than 50 years old)
- Male and female
- All people login to fill the questionnaire until we reached the required sample size.
- People filled out a consent form to participate in the study.

Exclusion criteria:

- People whose age was less than the selected age groups.
- People who did not meet the criteria of admission.

4. Sample size and technique:

Our sample was:

- The prevalence of SCD in Saudi Arabia is high, about 2% to 27% have sickle cell trait and 2.6% have SCD, so sample calculated was 410.
- Sampling: conventional sample.
- Selection of sample: when sample size had been completed, the application form closed.
- Sample size was estimated using EPI INFO (Epidemiological Information Package) version (21) 3.5.3.statistical packages assuming that the frequency was (20%) at a confidence interval of 95 % and power of 80%.

B-Operational design

1. Pilot study

A pilot study was carried out to evaluate the validity and reliability of the questionnaire applied on patients. Test-retest reliability was assessed using the questionnaire twice on 10% of the sample size (41subjects). Based on the result of the pilot study some modifications and rearrangement of some questions were done. Validation of the questionnaire was made as follows: the questionnaires were translated using a back-translation technique. An expert translated the original questionnaire from English into Arabic. Arabic version of the questionnaires was translated back into English by a bilingual individual. The back-translated and original versions of the questionnaire were compared with attention given to the meaning and grammar.

Data collection tools:

Tools:

Self-administered questionnaire divided into three sections (Boyd et al., 2005 and Alturaifi et al., 2018)17:

1-First Section:

Included personal and socio-demographic data:

Age, gender, marital status, educational level and occupation

2-Second section:

Included participant perception about SCD which was measured in 6 close-ended questions.

3-Third section:

Included Knowledge about SCD and its' complications, which was measured by 11 closed-ended questions.

Knowledge score: Included knowledge of the studied participants about SCD. A total Knowledge score was calculated, eight questions were given one point for answer yes, and zero for answer no, or I don't know, while four questions were calculated as each correct answer or the proper answer selection were given one point; summation of the correct answers for all questions was done and given 35 points as total knowledge score. So, the higher the score, the better the knowledge.

2. Data management:

The Collected data were recorded then presented and analyzed using SPSS (Statistical Package for the Social Sciences) version 22.0 and Epi info for windows version 3.5.3.

Data were represented in tables and graphs as frequencies and percentages.

C. Ethical considerations:

Ethical considerations were taken throughout the whole study including approval of the study protocol by Institutional Human Ethics Committee, Ibn Sina National College for Medical Studies, Jeddah, KSA; agreement of the participants to fill out the questionnaire was taken after explaining the purpose of the study and assuring them regarding data confidentiality, however, each subject was given a unique identifier code.

D. Constraints:

Some constraints were involved as we needed to increase the sample taken to avoid the not completely filled out questionnaires.

Results

The study sample included 410 participants most of them females (91%) and 9% males. More than 50% of participants were between the age group 20 to 30 years old and about 59.9% were married. 73.4% were university students (Table 1, Figures 1-5).

Table 1: Demographic characteristics among studied subjects

Demographic Character		No	%
Age	20-30	241	58.8
	31-40	78	19
	41-50	64	15.6
	>50	27	6.6
	Total	**410**	**100**
Gender	Female	375	91
	Male	37	9
	Total	**410**	**100.0**
Marital Status	Single	117	43.2
	Married	217	59.9
	Widow	11	2.7
	Divorced	5	1.2
	Total	**410**	**100.0**
Educational level	1ry school	2	0.5
	Intermediate school	9	2.2
	High school	81	19.8
	University	301	73.4
	High education	17	4.1
	Total	**410**	**100**
Occupation	Employee	145	35.4
	Un employee	48	11.7
	Student	133	32.4
	Housewife	84	20.5
	Total	**410**	**100**

Figure 1: Age distribution of studied subjects

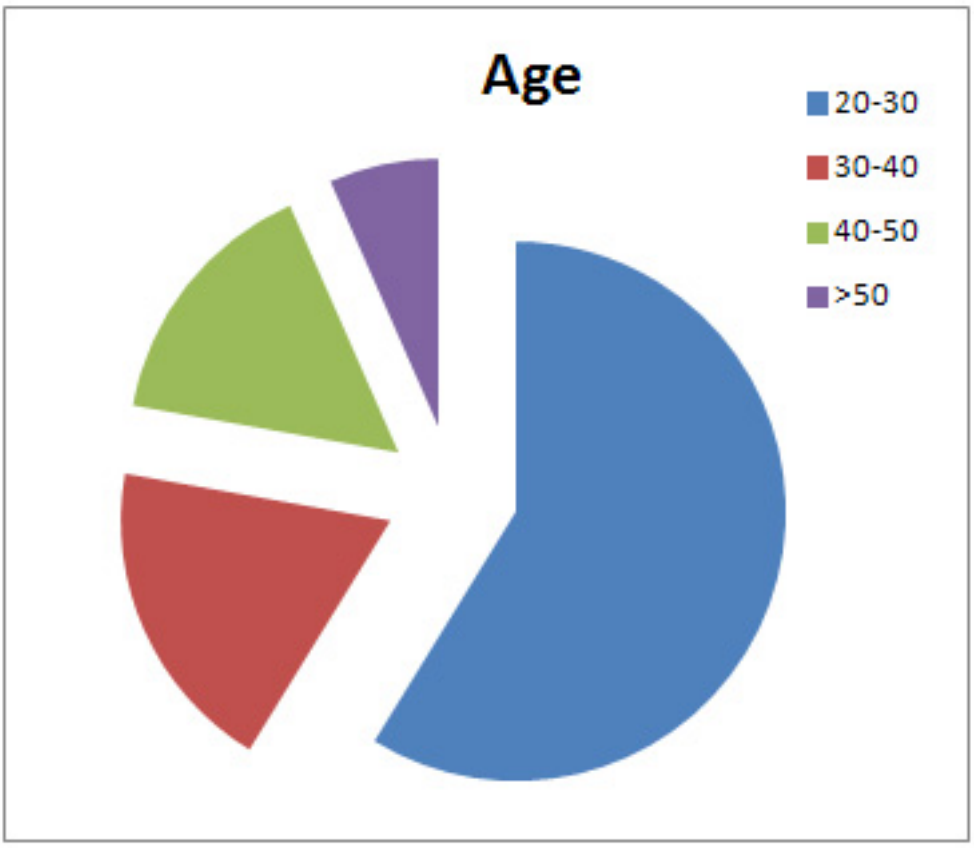


Figure 2: Gender distribution of studied subjects

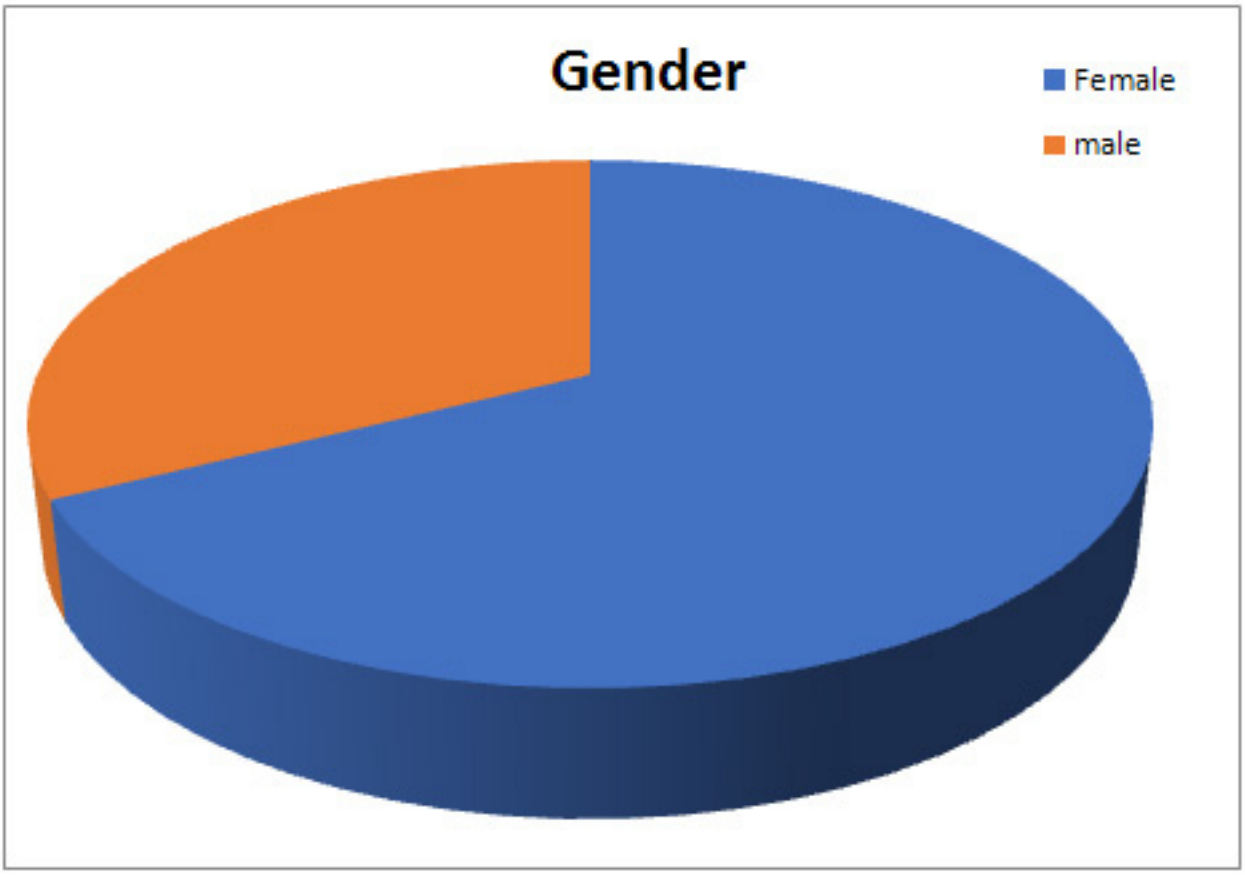


Figure 3: Marital status of studied subjects

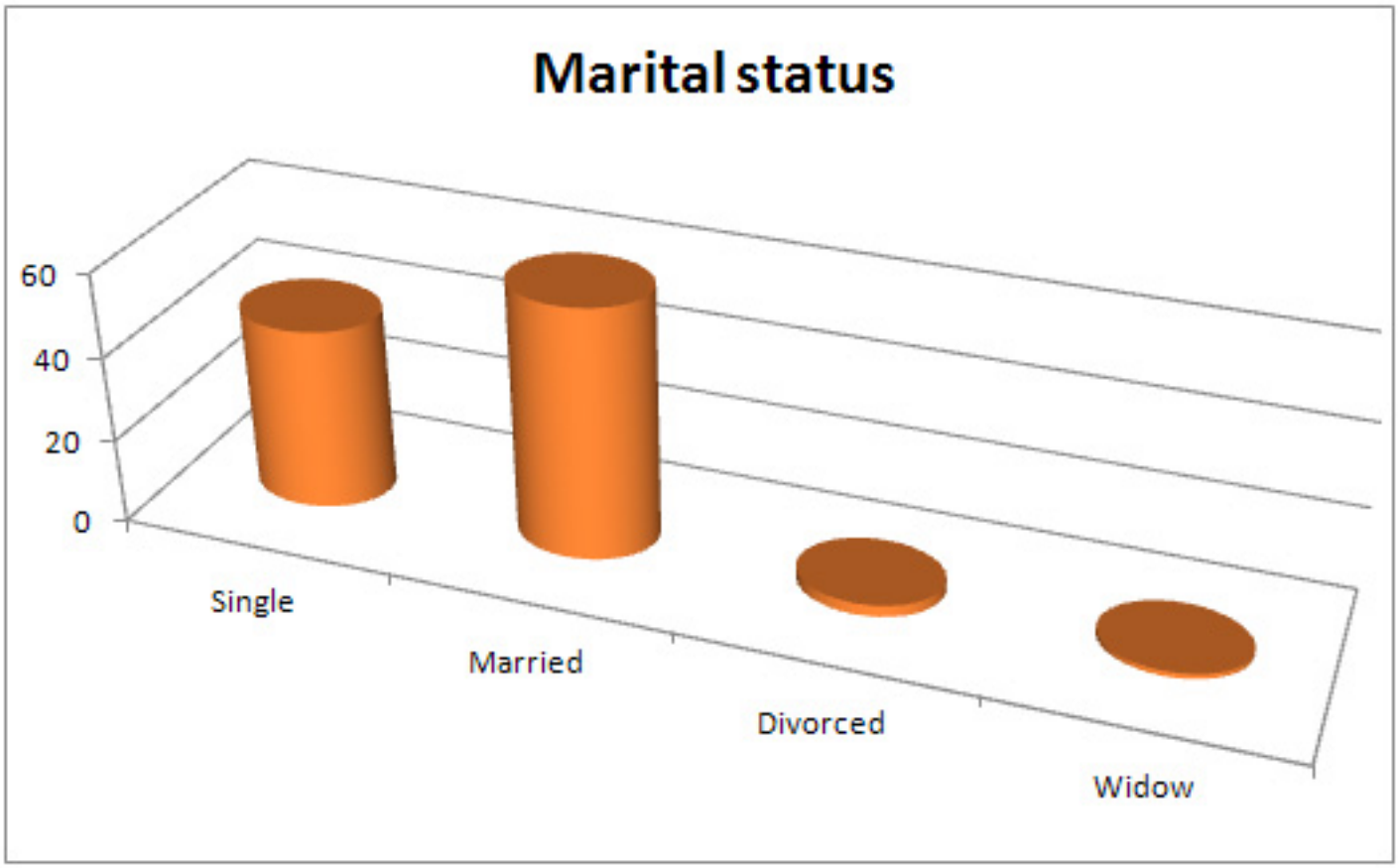


Figure 4: Educational level of studied subjects

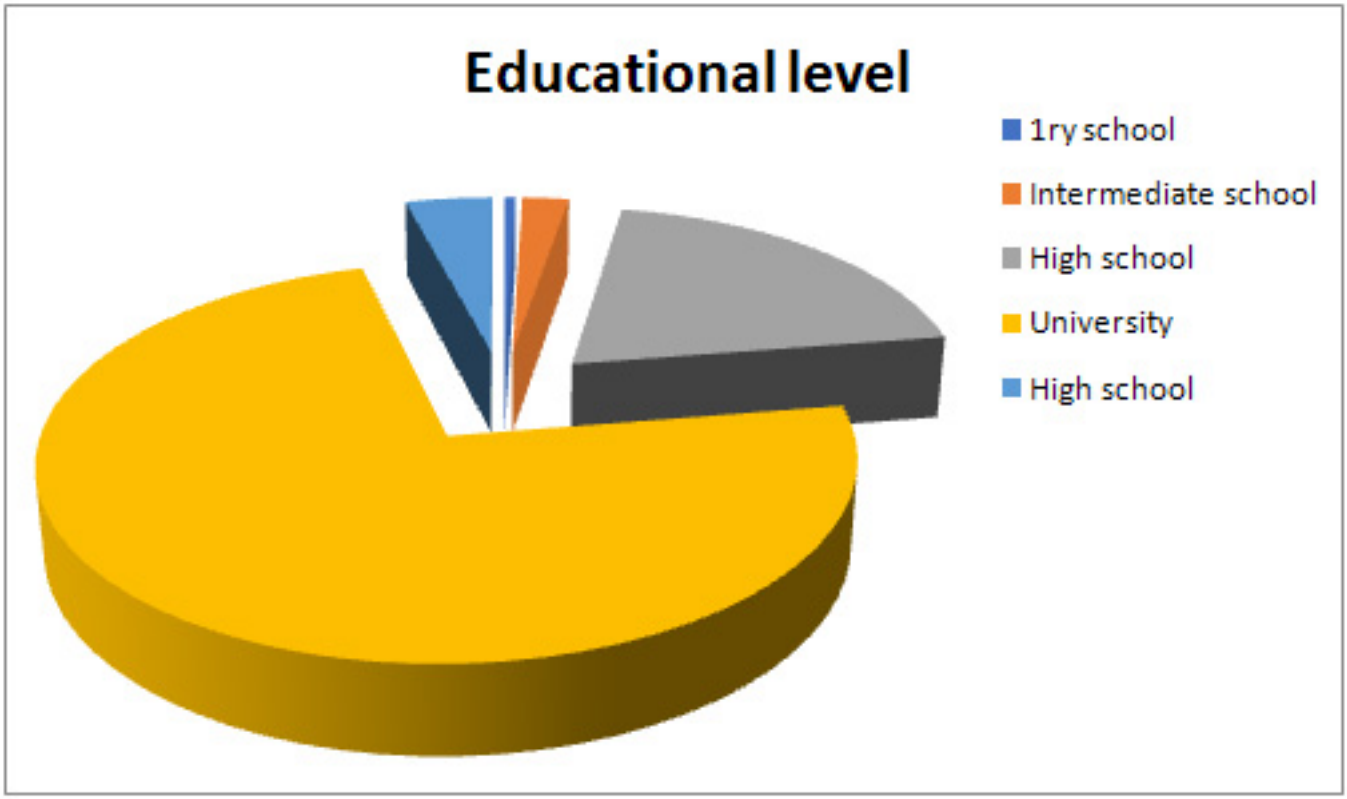
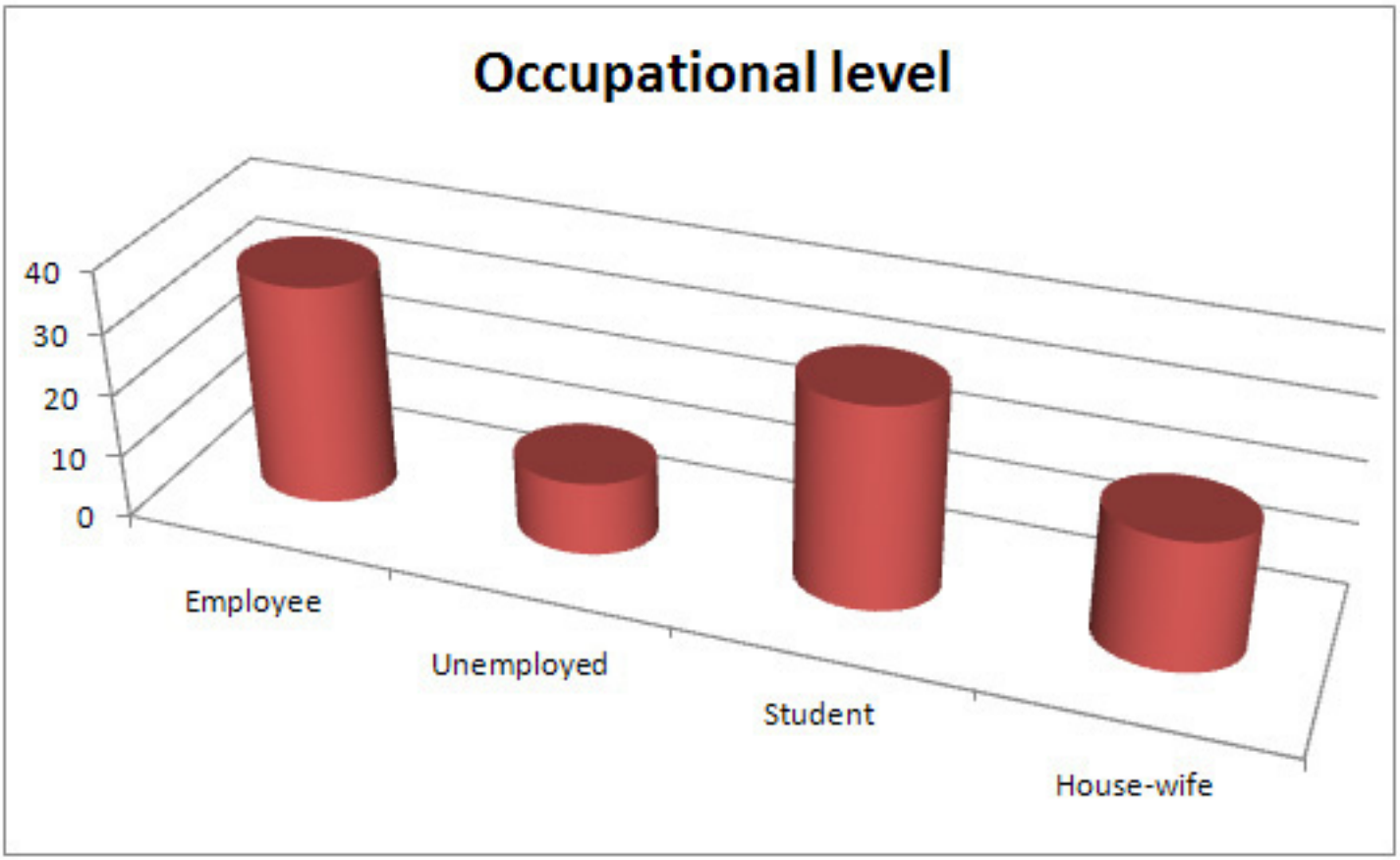


Figure 5: Occupational level of studied subjects

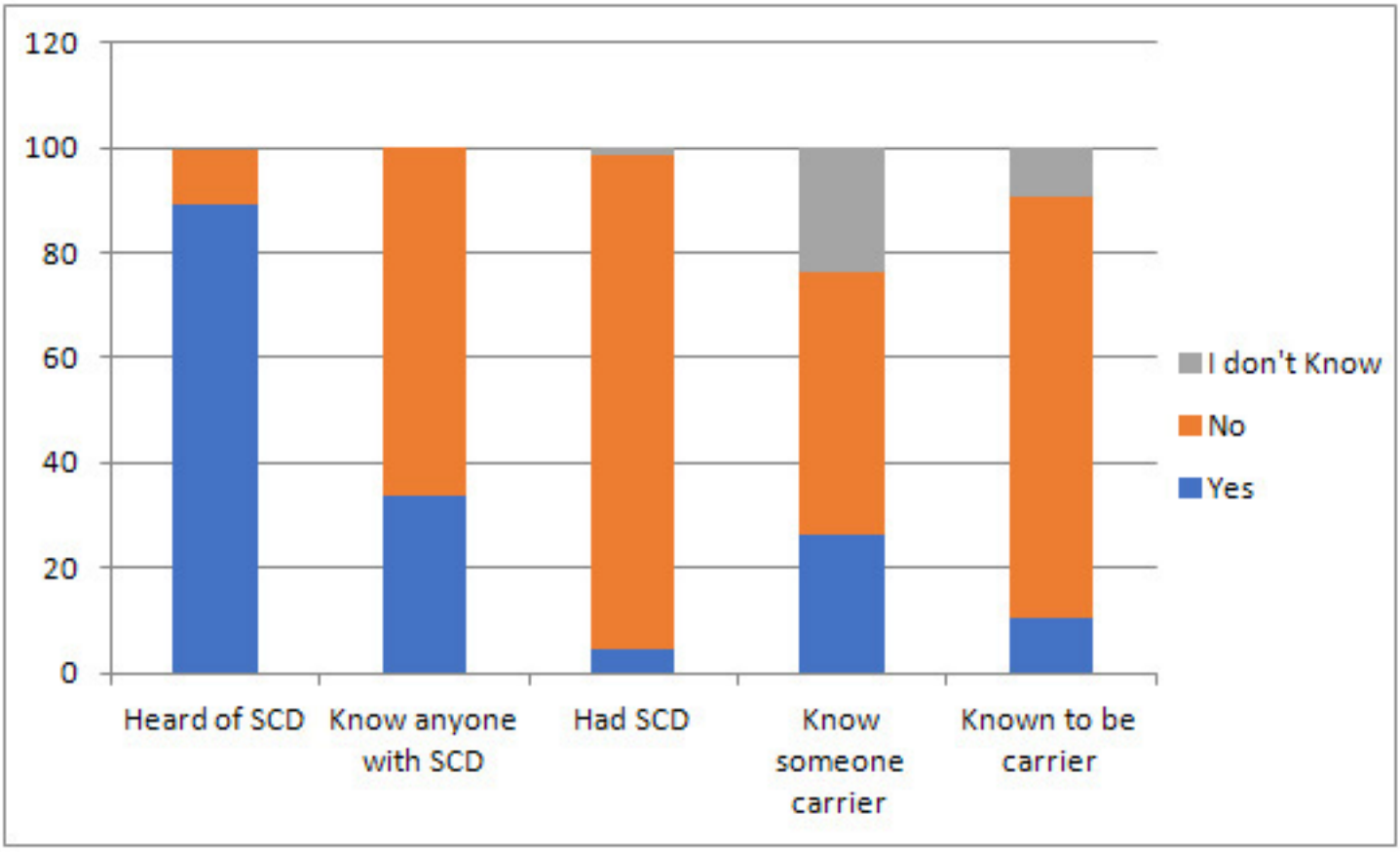


Perception of participants to sickle cell anemia, found that the majority of them (89.5%) had heard about the disease; 44.4% heard about it from the school. More than 66% of the participants did not know anyone with SCD, only 4.4% had the disease and 10.5 were known to be carrier as shown in Table 2.

Table 2: Participants' perception of Sickle cell disease

		No	%
Have you ever heard of Sickle cell disease?	Yes	365	89.5
	No	43	10.5
	Total	**408**	**99.5**
How did you hear about it?	Awareness program	156	38
	Healthcare worker	71	17.3
	At school	182	44.4
	Total	**409**	**99.8**
Do you personally know anyone with sickle cell disease?	Yes	139	33.9
	No	271	66.1
	Total	**410**	**100.0**
Do you have sickle cell disease?	Yes	18	4.4
	No	387	94.4
	I don't know	5	1.2
	Total	**410**	**100**
Do you know anyone who is sickle cell carrier?	Yes	108	26.3
	No	204	49.8
	I don't know	98	23.9
	Total	**410**	**100**
Are you a sickle cell carrier?	Yes	43	10.5
	No	328	80
	I don't know	39	9.5
	Total	**410**	**100**

Figure 6: Bar-chart showed participants perception of SCD



Knowledge of participants about cause of sickle cell disease was measured; we found that more than 63% of participants know that it is an inherited disorder and more than half of participants had information that the disease runs in generations but can skip a generation sometimes. Regarding women with SCD who can get pregnant or not and if it affects the pregnancy and fetus, more than 60% of participants answered that a woman can get pregnant and nearly the same percent found that it affects the pregnancy (Table 3 & Figure 7).

Figure 7: Bar-chart showing participants knowledge about SCD

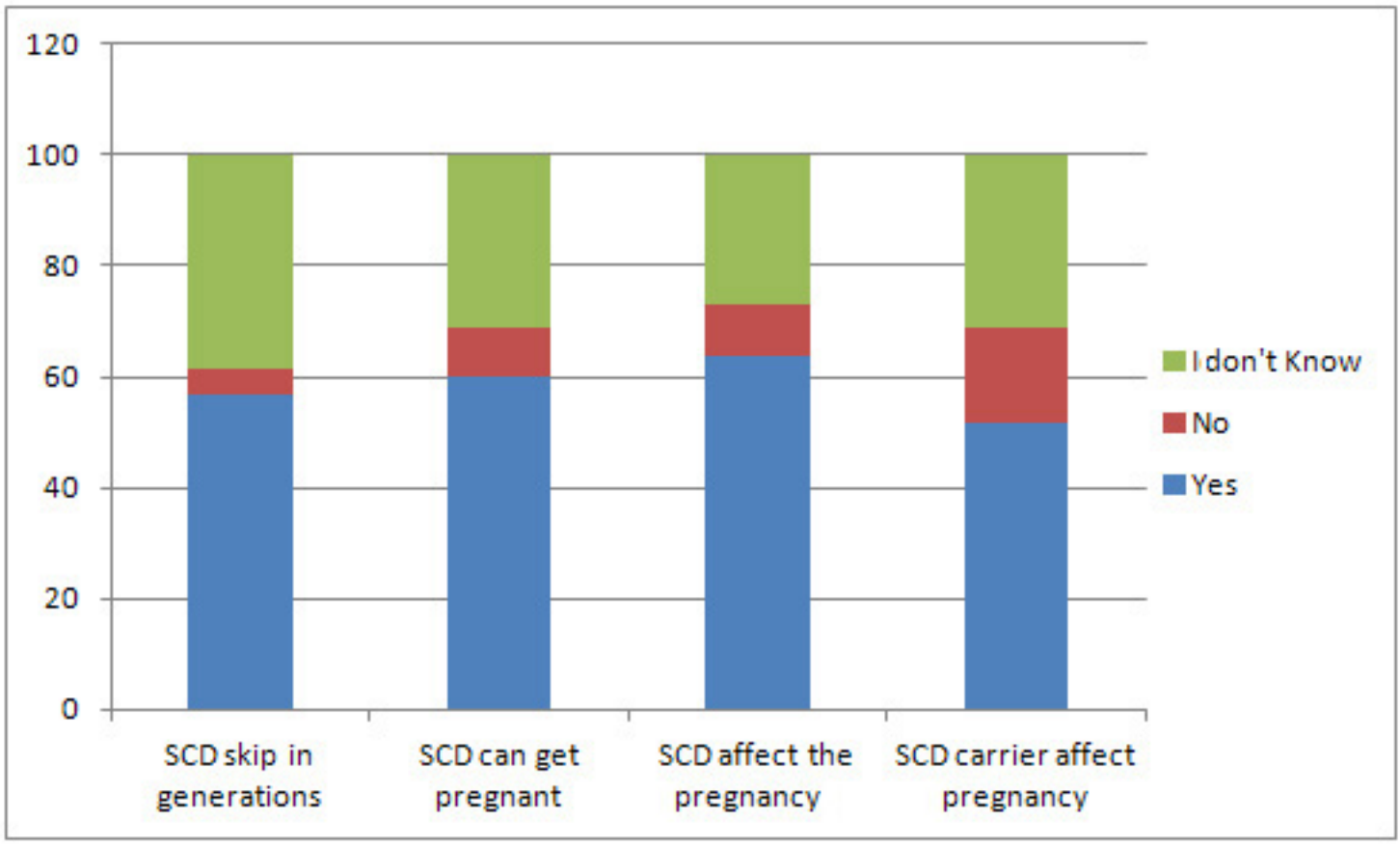


Table 3: Information of the studied subjects about inherited sickle cell disease

		No	%
How do you get Sickle cell disease?	Hereditary	259	63.2
	Acquired	41	10
	I don't know	110	26.8
	Total	**408**	**99.5**
Does Sickle cell disease sometimes skip generations in families?	Yes	233	56.8
	No	19	4.6
	I don't know	158	38.5
	Total	**409**	**99.8**
Do you think that women with sickle cell can get pregnant?	Yes	247	60.2
	No	36	8.8
	I don't know	127	31
	Total	**410**	**100.0**
Do you think that (sickle cell disease) affects the pregnant women?	Yes	261	63.7
	No	39	9.5
	I don't know	110	26.8
	Total	**410**	**100**
Do you think that (sickle cell trait) affects the pregnant women?	Yes	211	51.5
	No	71	17.3
	I don't know	128	31.2
	Total	**410**	**100**

Regarding participants' knowledge about sickle cell disease and its complications and how the pregnancy affects SCD, about 67.8% had previous experience that it affects the pregnancy by hematological complications, 23.4% by infection and 29.3% by the complication of blood transfusion. About the effect of SCD in pregnancy, 44.9% said that it may cause antepartum hemorrhage, 38% found it may affect fetus by causing intrauterine gross retardation (Table 4)

Table 4: Information of the studied subjects about sickle cell disease and its complications

		No	%
Which of the following are true of Sickle cell disease?	A blood disease	312	76.1
	Many types	92	22.4
	Identified by blood test	172	42
	Blood transfusion needed for treatment	120	29.3
How does the pregnancy affect the sickle cell disease?	Hematological complication	278	67.8
	Infection	96	23.4
	Blood complication	120	29.3
	Others	107	26.1
How does the sickle cell disease affect the pregnancy?	APH	184	44.9
	Gestational diabetes	49	12
	IUGR	156	38
	Eclampsia	83	20.2
	Pregnancy induced HPT	123	30
	Others	128	31.2

Regarding the participants' knowledge about how to deal with SCD patients, more than 76% found that they need plan for management and more than 80% said that they need special care during pregnancy and during delivery as investigation, regular follow-up, some medications and vaccinations, (Table 5).

Table 5: Information of the studied subjects about management of SCD

		No	%
Is it important to the affected women to plan and discuss the future pregnancy with the hematologist?	Yes	312	76.1
	No	19	4.6
	I don't know	79	19.3
	Total	**410**	**100**
Do you think pregnant women with sickle cell needs special care during her pregnancy?	Yes	347	84.6
	No	15	3.7
	I don't know	48	11.7
	Total	**410**	**100.0**
Do you think that if there is a special management done to the pregnant women with sickle cell during delivery?	Yes	358	87.3
	No	10	2.4
	I don't know	42	10.2
	Total	**410**	**100**

Regarding detection of the level of knowledge which was measured through the scoring applied we found that most of the participants had moderate knowledge about SCD (>60%) and there was no difference regarding socio-demographic characters.

Table 6: Association between levels of knowledge about sickle cell disease and Socio-demographic characteristics of the studied subjects

		Knowledge response		P-value
		Poor score	Good score <=35	
Age	20-30	200	41	0.681
	30-40	54	24	
	40-50	48	16	
	>50	20	7	
Gender	Female	220	155	0.782
	Male	30	7	
Marital Status	Single	89	28	0.230
	Married	165	53	
	Widow	8	3	
	Divorced	5	0	
Educational level	1ry school	2	0	0.681
	Intermediate school	7	2	
	High school	60	21	
	University	198	103	
	High education	10	7	
Occupation	Employee	104	41	0.361
	Un employee	35	13	
	Student	88	45	
	Housewife	64	20	

Discussion

SCD is a hemolytic anemia characterized by abnormally shaped (sickle) RBCs, which are removed from the circulation and destroyed at increased rates leading to anemia (1).

Pregnancy in SCD is at very high risk (14). Major steps for prevention is to carry out various programs, surveys, educating and increasing awareness toward the disease and its consequences including morbidity and mortality (11).

So, the aim of current study was to assess the awareness about complications of SCD during pregnancy in Jeddah city, Saudi Arabia.

The study sample included 410 participants, most of them female (91%) and 9% male. More than 50% of participants were aged between 20 to 30 years old and in about 59.9% their marital status was married. 73.4% were university students (Table 1, Figures 1-5).

Regarding perception of participants about SCD, it was found that the majority of them (89.5%) had heard about the disease, 44.4% heard about it from school. More than 66% of the participants did not know anyone with SCD, only 4.4% had the disease and 10.5% were known to be carrier as shown in Table 2 and Figure 6.

This is in agreement with the study conducted by Alturaifi et al., in which most (86.3%) of the study population had heard about SCD, and only 16 (3.8%) had previous experience with an SCD child (17).

Knowledge of participants about the cause of SCD was measured, and it was found that more than 63% of participants knew that it is an inherited disorder and more than half of the participants had information that the disease

runs in generations but can skip a generation sometimes. Regarding if women with SCD can get pregnant or not and if it affects the pregnancy and fetus, more than 60% of participants answered that a woman can get pregnant and nearly the same percentage found that it affects the pregnancy (Table 3 & Figure 7).

The above mentioned results were matched with the study performed by Alturaifi et al., who showed that most (77.8%) of the surveyed subjects knew that SCD is a hereditary disorder, and 254 (59.9%) subjects recognized that SCD sometimes skip generations in families (17).

Regarding participants' knowledge about sickle cell disease and its complications and how pregnancy affects SCD, about 67.8% had previous experience that it affects the pregnancy by hematological complications, 23.4% by infection and 29.3% by the complication of blood transfusion. About the effect of SCD in pregnancy, 44.9% said that it may cause antepartum hemorrhage, 38% found it may affect the fetus by causing intrauterine gross retardation (Table 4).

Similarly, Alturaifi et al., found that most (55.7%) of the respondents knew that SCD is a blood disease, and 74 (17.5%) stated that it could be identified by a blood test (17).

Also, it is consistent with the study conducted by Obed et al., who found that patients were most likely to answer correctly that SCD is a "blood disease" (130/206, 63.1%) (18).

Regarding the participants' knowledge about how to deal with SCD patients, more than 76% found that they need a management plan and more than 80% said that they need special care during pregnancy and during delivery such as investigation, regular follow-up, some medications and vaccinations, (Table 5).

Regarding detection of the level of knowledge which was measured through the scoring applied it was found that most of the participants had moderate knowledge about SCD (>60%) and there was no difference regarding socio-demographic characteristics (Table 6).

Our findings agreed with the results of Alturaifi et al., in which 51.4% of participants showed a good level of knowledge. However, our findings were in disagreement with the same study regarding the relationship between socio-demographic factors and level of knowledge where sex and education level had significant association with the level of knowledge ($p<0.05$) (17)

Consistent with a study in Bahrain by Al Arrayed and Al Hajeri, findings showed good level of knowledge about SCD among the public, while, findings were in contrast with them regarding the significant association of sex and education level with the level of knowledge (19).

In addition, Treadwell et al., reported that 68% of their study population responded correctly to knowledge questions about SCD(20).

In contrast, a low level of knowledge was reported among SCD patients in Al-Qatif area, Eastern Province, Saudi Arabia and secondary school students in Nigeria (21).

Furthermore, Siddiqui et al., revealed substantial knowledge gaps about sickle cells in surveyed people of reproductive age from the Dominican and African American communities in Northern Manhattan (22).

Conclusion

Overall knowledge about SCD and its complications during pregnancy was moderate. So, we recommend that health education programs about all aspects of SCD should be designed, implemented and evaluated among the general population.

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