

Knowledge, Attitude and Practices Related to Pre-Marital Screening for Sickle Cell Anemia in Jazan Region, Saudi Arabia

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Abstract

Background: Sickle Cell Anemia (SCA) is considered relatively common in the Kingdom of Saudi Arabia, particularly in the Eastern and Southwestern parts of the country. Previous studies showed significant widespread of misconceptions and poor knowledge about sickle cell disease in the Eastern region; however, data from the Southwestern areas are limited. **Objective:** Therefore, the present study aims to assess the knowledge, attitude and practices related to pre-marital screening for SCA in Jazan region. **Methods:** A cross-sectional study was carried out among 230 male Saudis who had been married in 2004 or after with positive SCA test for either partner or both. The participants were interviewed to fill up structured questionnaires at five public health centers (PHCs) in Jazan region (3 urban and 2 rural PHCs). **Results:** Overall, an average level of knowledge about SCD was reported among the participants, with 17.4% of them had poor knowledge. About half of the participants had the knowledge from their friends and neighbours. Interestingly, about 60% of the participants supported consanguinity, with about two-thirds had consanguineous marriage. Almost all (95.2%) of the respondents were in favour of performing screening for SCA and 83.5% of them preferred to do it before engagement. Participants with at-risk marriages had significantly higher level of knowledge compared to those with safe marriages ($\chi^2 = 9.19$, $P = 0.010$). Similarly, consanguineous marriage increased the odds of at-risk marriages by four times (OR = 3.7, 95% CI = 1.9, 7.0). **Conclusion:** The participants had an average level of knowledge about SCD. However, percentages of consanguineous marriages and completing at-risk marriages were unexpectedly high. Thus, health education should play a significant role in preventing genetic diseases and this might be integrated in the curriculum of secondary and university levels. Screening for SCA before engagement will help significantly to avoid at-risk marriage and to reduce these diseases.

Keywords: *Sickle cell anemia, pre-marital screening, knowledge, attitude, Jazan, Saudi Arabia*

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Introduction

Sickle cell disease (SCD), an inherited blood disease, is an increasing global health problem, with over 350,000 infants

are born with SCA every year.^{1,2} The disease is characterized by a modification in the shape of the red blood cell from a

smooth, donut-shape into a crescent or half-moon shape that lack plasticity and may block small blood vessels and impair blood flow. This will lead to shortened red blood cells survival, and subsequent anemia, often called SCA. Overall, SCD is difficult to treat and causes significant morbidity and mortality that pose an economic burden on both family and country levels.³ Globally, the prevalence of sickle cell trait ranged between 2% and 30% across Africa and it is also common in the Eastern Mediterranean, Middle East, India and in South and Central America.^{4,5} In Saudi Arabia, prevalence of haemoglobinopathies such as SCD and thalassaemia is considered high, particularly in the eastern (the eastern region and Al-Ahsa) and southwestern (Jazan and Qunfudah areas) parts of the Kingdom.⁶⁻⁹ The prevalence of sickle cell trait ranged from 2% to 27%, while SCD was up to 1.2% in some areas.⁷ This high prevalence of haemoglobinopathies could be attributed to the consanguineous marriage rate that reach as high as 67% in KSA.^{10,11} A previous cross-sectional study found that Among 488,315 individuals who applied for a marriage license during February 2004 and January 2005, 4.2% had sickle cell trait while 0.26% had SCD.⁷ Moreover, the study showed that out of 207,333 screened couples, 4,444 (2.14%) had mismatched results and were declared as high-risk marriage. Unfortunately, about 90% of those couples had completed their marriages, despite the known high-risk status.⁷

The pre-marital screening and genetic counseling (PMSGC) program for hemoglobinopathies was initiated in Saudi Arabia in February 2004, with a main aim of reducing the prevalence of SCD and β -thalassaemia among the population through reduction of the number of at-risk

marriages.^{8,12} Subsequently, a study conducted in Arar (a northern border region) revealed that at-risk marriage rate was decreased by about 60% between 2004 and 2009.¹³ In the same vein, a retrospective study reported a 5-fold increase in the number of voluntary cancellation of at-risk marriage proposals during the same period leading to constant prevalence of SCD in the country.¹⁴

Despite the fact that Jazan region ranks second in SCD after Al-Ahsa region⁷, data on people's knowledge about the disease and pre-marital screening are lacking. Thus, the present study aims to evaluate the knowledge, attitude and practices (KAP) towards the pre-marital screening program among newly married Saudis in Jazan region. It is hoped that findings of our study will be useful for health authorities to identify the underlying factors for the unchanged rate of high-risk marriages in Jazan. Such data will help to identify and implement integrated and effective preventive measures to reduce occurrence of high-risk marriages and to curtail haemoglobinopathies among this population.

Methods

Study setting: A cross-sectional survey was carried out at primary health care centers (PHCs) in Jazan region during October 2015. Jazan region, located in the southwestern part of the kingdom, about 1,200 km from Riyadh, is among the smallest administrative areas of the country, with an area of 11,671 km² and a population of about 1.4 million.¹⁵ The region, comprises 14 governorates, is located alongside the Red Sea from its western borders with over 100 islands located in the sea and a 120-km border with Yemen at the south. Jazan is the capital city of the region; it involves one of important ports for the Kingdom.

Study population: A convenience sample of 230 Saudi male adult individuals identified in 5 primary health care centers (3 urban and 2 rural) in Jazan province, during October 2015 was considered for this study. The study participants were identified through screening the adult male attendees of the five primary health care centers for Saudi male adult individuals who fulfil the following two criteria: Had married during the year 2004 (the year of start of premarital screening program in Saudi Arabia) or after, positive premarital screening SCA result for the individual or his partner or both.

The study protocol was approved by the Ethical Committee of the Faculty of Public Health and Tropical Medicine, Jazan University, Jazan, KSA. A clear description about the objective of the study was given to the participants before obtaining their consents and commencing the interview. The participants were informed that their identity and personal data would be kept strictly confidential, and their participation was voluntary.

Questionnaire: A structured questionnaire was used to collect data on the demographic factors, KAP data, and pre-marital screening test results. The participants were face-to-face interviewed by trained research assistants at the respective PHCs. The questionnaire involved ten questions related to the knowledge about the pre-marital screening program. Respondents' answers of these questions were then recorded as scores (a score of "1" was given for the correct answer and a score of "0" was assigned to the wrong answer). "I do not know" answer was also treated as incorrect and given a score of "0". The ten question scores of each individual questionnaire were summed up to get a total score out of

ten for each respondent. The respondents' knowledge was classified according to the total scores into three categories; poor knowledge (scores from zero to four), average knowledge (scores from five to seven), and good knowledge (scores from eight to ten).¹⁶

The attitude questions were about favor of the consanguinity and support of the SCA premarital screening test. The practice questions were related to decision taken by the individuals and their partners in response to the premarital SCA positive result, in terms of cancellation or continuing the marriage proposal and whether their wives are from their relatives.

Data analysis: Data were double-entered into Microsoft Excel 2010 spreadsheets and were cross-checked for accuracy by another researcher before it was exported to SPSS, version 20 (IBM Corporation, NY, USA) for data analysis. For descriptive analysis, proportion was used to present the distribution of factors. Pearson Chi square test was used to assess the difference of proportions between groups while t-test was used to assess the difference of mean scores. Odds ratios (OR) and corresponding 95% confidence intervals (CI) were calculated. A *P* value of < 0.05 was considered statistically significant.

Results

A total of 230 Saudi males aged between 19 and 47 years with mean age of 30.2 years (SD 4.6) were enrolled in the study. Table 1 shows SCA pre-marital screening test results for the study participants and their partners. Overall, 150 (65.2%) participants had safe marriages with no risk of SCA to their children while 80 (34.8%) of participants had risky marriages due to incompatible results.

Table 1: Distribution of participants according to pre-marital screening test results

Characteristics	N	%
SCA test results for study participants & their partners		
Healthy and diseased	28	12.2
Healthy and carrier	122	53.0
Carrier and diseased	18	7.8
Carrier and carrier	57	24.8
Diseased and diseased	5	2.2
Marriage category		
Safe	150	65.2
At-risk	80	34.8

Table 2: Knowledge of the participants about SCA

No.	Knowledge statement/question	Correct answer (%)
1	Do you think consanguinity increases the probability of SCD?	72.6
2	Do you think that SCA carrier has symptoms?	55.2
3	Do you think there is a risk SCD to children of diseased couples?	90.4
4	Do you think there is a risk of SCD to children of carrier couples?	73.0
5	Do you think there is a risk of SCD to children of marriage between a healthy and a carrier?	79.1
6	Do you think that SCA carrier should abstain marriage in order not to have diseased children?	79.6
7	Do you think that SCD patient should abstain marriage in order not to have diseased children?	66.1
8	Do you think that blood transfusion cures SCD completely?	59.6
9	Do you think there is a complete cure for SCD?	48.3
10	Do you think that SCA carrier can be converted to SCD Patient?	63.9

Table 3: Knowledge, attitude and practices of the participants towards SCA and consanguinity

Characteristics	Number	%
Knowledge about SCD		
Good	95	41.3
Average	95	41.3
Poor	17	17.4
Attitude towards pre-marital screening test		
Support	220	95.7
Do not support	10	4.3
Suitable time for pre-marital screening test		
Before engagement	192	83.5
After engagement	38	16.5
Favor consanguinity	137	59.6
Completing at-risk marriage	77	96.0
Had consanguineous marriage	146	63.5

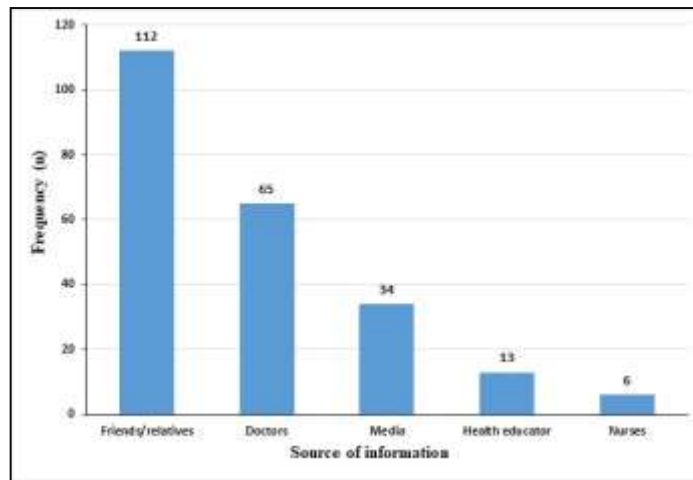


Figure 1: Source of information about SCD among the participants

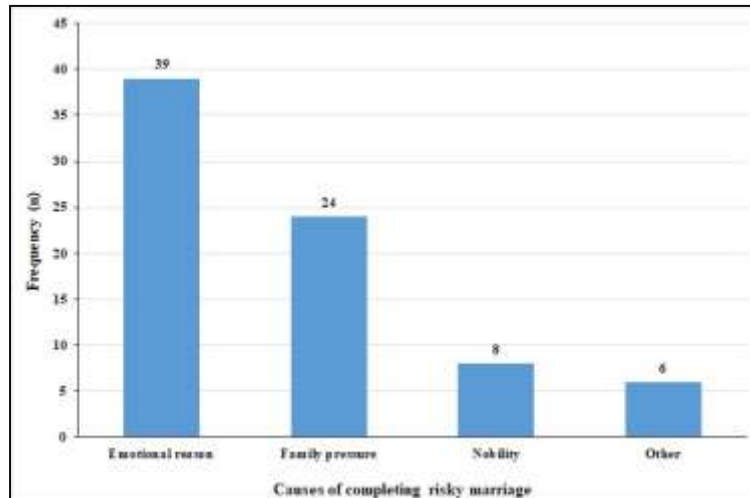


Figure 2: Causes of completing risky marriages among the participants

Participants’ knowledge attitude and practices towards SCD: Knowledge level of participants about SCA was assessed using arbitrary scoring system based on 10 questions (Table 2). The participant scores ranged between 2 and 10, with the mean score was 6.88 (SD = 2.19); indicating an average level of knowledge about SCA among the study population.

The results revealed that majority (82.6%) of the participants had average-to-good knowledge about the disease while 17.4% (40/320) of them had poor knowledge (Table 3). With regards to source of information, friends and relatives comprised the main source (48.7%) while all healthcare personnel (doctors, nurses and health educators) contributed by one third (36.5%) only (Figure 1).

Table 3 shows that almost all (95.2%) of the participants were in favour of performing screening for SCA and 83.5% of them prefer doing the test before engagement. The percentage of those who supported consanguinity was high (59.6%) and 63.5% (146/230) of the participants had married their relatives. With regards to their practices, the majority of participants (96%) completed their marriages proposal.

About half (50.6%) of them declared emotional reasons as the cause of completing the at-risk marriages while about one third (31.2%) of them had been pressured by their families (Figure 2).

Further analysis showed that participants whose their marriages were risky had significantly higher level of knowledge about SCA, represented by the mean scores, compared to those who had safe marriages (7.4±1.9 vs. 6.6±2.3, $t = 2.9, P = 0.004$). Accordingly, proportion of participants with good level of knowledge was significantly higher among those who had at-risk marriages compared to their counterparts (50.0% vs. 36.7%, $\chi^2 = 9.19, df = 1, P = 0.010$).

Similarly, significant association between consanguinity and at-risk marriages was found. The participants with consanguineous marriages were about four times more likely to had at-risk marriages than their non-consanguineous counterparts (44.5% vs. 17.9%, OR = 3.7, 95% CI = 1.9, 7.0).

Discussion

The present study was conducted among adult males attending PHCs in Jazan,

Saudi Arabia. The participants recruited to the study involved all possibilities of at-risk marriages, ranged from the least risky marriage (carrier + healthy) to the absolute risky marriage (diseased + diseased). Despite the convenience sampling strategy (recruited those who were positive for SCA; either one or both partners) by the present study, 17.4% of the participants had poor knowledge about SCA. However, the percentage of 82.6% of those who had average-to-good knowledge about SCA revealed the good impact of the Saudi pre-marital screening and genetic counseling (PMSGC) program. In comparison, a previous study in Saudi Arabia found fair level of knowledge about the nature of the pre-marital tests and the targeted diseases among different groups of participants including couples applying for PMSGC as well as those who had received the results of their screening.¹⁷

By contrast, a study among 655 attendees of governmental outpatient clinics in Jeddah reported low participants' knowledge about the PMSGC program.¹⁸ Regionally, about 83% of 590 unmarried Omani university students and 85% of 400 Omani adults who attended primary healthcare institutions had average-to-good knowledge about the pre-marital tests.^{19,20}

Similarly, Jordanian university students had good knowledge about the existence and nature of the program; however, about half of the respondents had a remarkable lack of knowledge about the type of diseases to be screened.²¹ A previous study among 800 students at King Abdulaziz University, Jeddah found that less one third of the students were able to give correct answer when questioned about the type of diseases screened by the PMSGC.²² The study also showed that information about this health problem was mainly acquired from friends and neighbours

(48.7%) while official and specialized sources represented only one third (36.5%). Thus, more efforts by the PHCs and related agencies are required to achieve the desired target.

Our findings showed that attitude of the participants towards consanguinity was consistent with their practices. The percentage of those who support consanguinity was about 60% and the percentage of those who already married from their relatives was 63.5%. It was estimated that many Arabian countries display some of the highest rates of consanguineous marriages in the world ranging from 20 to 50% of all marriages, with about 20-30% favouring first cousin marriages.^{10,23} Nonetheless, rate of consanguinity in Saudi Arabia ranges from 42% to 67%.^{18,24-27} In parallel, findings of the present study showed that consanguineous marriages were at 4-fold higher risk to had at-risk marriages than non-consanguineous marriages. Interestingly, a very much lower rate (14.5%) of consanguinity was reported among married adults who attended primary healthcare institutions in Oman, with 72.4% of the single participants preferred to avoid consanguineous marriage.²⁰

Although the participants with at risk marriage of SCA babies (80 individuals) knew that their planned marriages were risky, 96% of them had completed their marriages. This finding is slightly higher than that reported earlier.⁷ About 90% of 4,444 couples who had mismatched results from all over the Kingdom (193 centers and laboratories) had completed their at-risk marriages.⁷ Subsequently, another study reported increasing trend of the voluntary cancellation of either SCA or β -thalassaemia at-risk marriages, from 9% in 2004 to 52% in 2009 in the whole

kingdom.²⁸ That said, about half of couples with at-risk marriages complete their proposals in the general Saudi population while in Jazan region the percentage is still very high (96%) as reported by the present study. This variation could be due to the cultural and traditional differences between Jazan and other areas of the kingdom.

Investigating the reasons, the present study revealed that most of the participants declared emotional feelings towards their partners (50.6%) and family pressure (31.2%) as the causes of continuing at-risk marriage. As a Middle Eastern cultural custom, engagement cancellation is very unfavorable, as it would negatively affect the girl's future opportunities to get married.²¹ Moreover, we found that most of the participants (83.5%) preferred to do the pre-marital screening test before engagement. This finding is consistent with the result reported among Jazan University students.²⁹ Moreover, Omani university students showed more advanced attitude as 24% of them preferred to do the test in earlier periods such as on joining colleges or even in high schools.¹⁹ In Canada, such genetic screening is performed in high school.³⁰ It is believed that screening for haemoglobinopathies during young adult lives will ensure to gain the maximum benefit of the screening and counseling programs. Moreover, knowing the status before engagement will diminish the impact of self-emotions and family pressure towards completing at-risk marriages.

We acknowledge some limitations that should be borne in mind when interpreting the findings of this study. First, this study had recruited male participants only; previous studies have showed significantly higher levels of knowledge about the pre-marital testing among females compared to males.^{21,31} This was attributed to that;

females are more concerned about the prevention of inherited diseases that may affect their babies. However, other studies found no significant difference in the level of knowledge according to gender.^{18,32} Second, findings on the consanguinity rate might be not applicable to be used to estimate the rate in the population because the participants were not drawn randomly from the general population and all of them had positive SCA tests (either one or both partners).

In conclusion, the present study revealed that the study participants had an average level of knowledge about SCA (scoring 6.9 out of 10), with 17.4% of them had poor knowledge. The main source of information about SCA was friends and neighbours. The percentage of those who supported consanguinity was unexpectedly high (60%); however, majority (83.5%) of the participants preferred doing the screening test before the engagement. The study also revealed that 96% of those marriages classified as at-risk marriages had completed marriage proposal, with main reasons: emotional feelings towards the other partner (50.7%), family pressure to complete marriage proposal (31.2%). Thus, health education should play a significant role in preventing high risk marriages and this might be integrated in the curriculum of secondary and university levels. Optimum timing for SCA test could be reconsidered; knowing their status before engagement will help significantly in reducing this problem.

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Competing Interests

The author declares no conflict of interest.

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