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RESEARCH ARTICLE

ASSESSING PREMARITAL CARRIER SCREENING AND KNOWLEDGE ABOUT SICKLE CELL DISEASE AMONG UNIVERSITY STUDENTS IN OMAN

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ABSTRACT

Background: Sickle cell disease (SCD) is a group of genetic diseases which is especially prevalent in tropical and subtropical regions. Pre-marital carrier screening is imperative to control this huge burden of morbidity and mortality. The aim of this study is therefore to assess the level of awareness, knowledge and attitude about Sickle Cell Disease and pre-marital carrier screening among undergraduate students. **Methods:** This cross-sectional, descriptive study was conducted with undergraduate medical and pharmacy students studying in the National University Science and Technology. Ethical approvals were obtained from institutional review committees. Using non-probability sampling technique, study participants were recruited from National University Science and Technology. Data collection was carried out using a structured questionnaire. Statistical analysis was performed using SPSS (IBM SPSS Statistics 24.0). **Results:** Two hundred and seventy nine (n=279) participants who met the study's inclusion criteria were participated, of which 27 (9.7%) were males and 252 (90.3%) were females. Majority of study participants (81.4%) were aged between 20-25 years old and single (92.1%). Fifteen percent of the participants were undergraduate pharmacy student and about 85% were medical students. Less than one third (29.1%) of students were tested for SCD earlier, of which 3.8% had SCD and 22.8 had sickle cell trait. Majority were aware of benefits knowing the SCD status in both partners. One third thinks it is hard to convince the partner for screening and 40.5% do not support consanguineous marriage. **Conclusion:** Medical and pharmacy students have adequate knowledge regarding SCD, however the practices and attitude is not appropriate. The development of multifaceted patient and public health education and screening for the control of SCD by heterozygote detection, particularly during routine premarital medical examinations is needed.

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INTRODUCTION

Sickle cell disease (SCD) is one of the most common genetic disorders of hemoglobin worldwide (Oman Hereditary Blood Disorder Association, 2017). Sickle hemoglobin (HbS) is the most common structural mutation of normal adult hemoglobin (HbA), which is inherited as a Mendelian trait. Heterozygous carriers who inherit one HbS allele and one HbA allele are usually asymptomatic; in contrast, homozygous carriers who inherit HbS alleles from both parents suffer from SCD, which often causes intermittent vaso-occlusive crises resulting in tissue ischemia, acute and chronic pain and consecutively organ damage (WHO, 2011). It is estimated that each year over 332 000 affected births happen, about 275 000 have a sickle-cell disorder, with the majority in low and middle income countries , approximately 5% of the world's population are healthy carriers of a gene for sickle-cell disease or thalassemia (Piel, 2010). Children and adults born with SCD in the developed world had longer lives with fewer complications, due to early diagnosis and the availability of treatment options (Rees, 2010). There is high effect of genetic

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blood disorders in Arab population due to high consanguinity marriage rate (20-60%) (Al-Riyami, 2003). Annually 6491 conceptions are affected with Sickle Cell Disorders in WHO Designated Regions of Eastern Mediterranean region. The likelihood of children being born with a major haemoglobinopathy in Oman would be about 3 per 1,000 births (Al-Riyami, 2001). The reported prevalence of sickle cell trait was 6% and β-thalassemia trait was 2%, whereas the prevalence of homozygous sickle cell and β-thalassemia was 0.2% and 0.07% respectively. According to the last field study of Oman Hereditary Blood Disorder Association, 6% of children under 5 years old are carriers of a gene for blood disorders, while the disease accounts for 0.2% of the population and it is more in North Al-Sharqiya, Al-Dakhiliya, Muscat, South Al-Batinah and Mussandam (Rajab, 2013). Although Omani society is multi-ethnic, the prevalence of consanguineous marriages is 58% (Wright, 2010). All of these genetic disorders are significant burden on healthcare systems. Their chronic nature requires life-long medical attention and expensive therapy and specialized care. For these reasons, the premarital screening (PMS) was introduced in Oman in 1999. One way to prevent hereditary diseases is the premarital carrier screening (PMCS) service, it covers screening for the most common haemoglobinopathy such as sickle cell disease (SCD) and thalassemia and available in all health centers, individuals receive health education about hereditary disorders too [9]. Due to the high prevalence of sickle cell disease and its impact in the Omani society, it is important to find the knowledge and awareness level among Omanis. And on the basis of these statistics and data awareness campaign and prevention plan can be designed to decrease the disease burden. Multiple published studies have given varying reports about the knowledge of sickle cell disease among students, but there is paucity of published data on the knowledge of sickle cell disease among students in our locality (Adewuyi, 2000). Knowledge about Sickle Cell Disease is a way of preventing and controlling the scourge, since people will be better equipped to take informed decision concerning their marriage and the youths are good entry point for interventions aimed at controlling the disease. Since tertiary educational institutions in the country have good representation of Omani youths, there is need to assess the level of knowledge, ignorance and misconceptions about sickle cell disease because an understanding of these factors will help to fashion appropriate public health education programs to increase awareness and knowledge of the condition. The aim of this study is therefore to assess the level of awareness, knowledge and attitude about Sickle Cell Disease among undergraduate students of medical and Pharmacy College.

MATERIALS AND METHODS

This is a descriptive cross-sectional study was conducted on medical and pharmacy students at National University Science and Technology. Data collection was carried out using a structured survey questionnaire, especially designed for this study. Survey instrument was developed based on literature reviewed and brain storming sessions by the research team members. Study questionnaire included socio-demographic characteristics, knowledge about sickle cell disease including prevalence, misconceptions, cause and problems associated with sickle cell disease and methods of prevention. During data collection two research assistants who were adequately trained were available for explanation and retrieval of the questionnaire. Informed consent was obtained from each of the students before the study commenced and after explaining the rationale for the study and ethics to them as a group. Sample size for this study was estimated using this formula for cross-sectional surveys: The required sample size was calculated by RaoSoft sample size calculator by means of prevalence of knowledge gap about Sickle cell disease among students, which is 73.4%, as reported by a previous study (Durotoye et al. 2013). The calculated sample size was 296. The minimum sample size (n) for this study was estimated to be 296. Likert-Scale was utilized for study questionnaire, where the participants were either strongly disagree, disagree, neither agree nor disagree, agree, or strongly agree with a number of statements regarding health beliefs related sickle cell disease. A data analysis was conducted using SPSS (IBM SPSS Statistics 24.0). Descriptive statistics analyses were performed for the study categorical variables. Data was expressed in frequencies for questionnaire responses calculated for all variables in numbers and percentages. Mann-Whitney test was used to compare differences between two groups with ordinal data and independent sample t-test was used to compare group differences with interval data.

RESULTS

Two hundred and seventy nine (n=279) participants who met the study's inclusion criteria were participated, of which 27 (9.7%) were males and 252 (90.3%) were females. Majority of study participants (81.4%) were aged between 20-25 years old and single (92.1%). Fifteen percent of the participants were undergraduate pharmacy student and about 85% were medical students. Less than one third (29.1%) of students were tested for SCD earlier, of which 3.8% had SCD and 22.8 had sickle cell trail (Table 1).

Table 1. Study Participants selected Demographics (n-279)

	Frequency	Percentages
Age		
<20 years	39	14.0
20-25 years	227	81.4
>25 years	13	4.7
Gender		
Male	27	9.7
Female	252	90.3
Marital Status		
Single	257	92.1
Married	21	7.5
Widow	1	0.4
Area of Study		
Medicine	238	85.3
Pharmacy	41	14.7
Tested for SCD earlier?		
Yes	81	29.1
No	158	56.8
Don't know	39	14.0
If yes, what was the test result?		
I have sickle cell disease	3	3.8
I have sickle cell trait	18	22.8
I don't have sickle cell disease	56	70.9
I don't know	2	2.5

Series of questions were asked from study participants regarding their knowledge about sickle cell disease (Table 2). Majority of study participants (98%) were aware of SCD and more than half think prevalence of SCD in Oman is between 10-50%. About 61% think SCD treated by bone marrow transplantation and 25% believed via blood transfusion.93% agreed that SCD is a disease of genetic or inheritance. GPs were asked multiple questions regarding SCD and Marriages practices.

Table 2. Study Participants Knowledge about Sickle Cell Disease

	Frequency	Percentages
Have you ever heard about SCD before?		
Yes	273	97.8
No	6	2.2
Do you know how prevalent SCD is in Oman?		
Yes	180	64.5
No	99	35.5
If you answered yes, what do you think the percentage is approximately?		
<10%	15	8.4
10-50%	121	67.6
>50%	43	24.0
Parents with SCD, chances of having children with SCD?		
0%	47	17.1
25%	132	48.0
50%	86	31.3
75%	7	2.5
100%	3	1.1
How is sickle cell disease treated		
Antibiotics	8	2.9
Liver transplantation	3	1.1
Rest	2	0.7
Bone marrow transplantation	170	60.9
Blood transfusion	69	24.7
None of the above	27	9.7
Cause of sickle cell disease		
Malnutrition	2	.7
Genetic/inherited	266	95.7
Infections	1	.4
Hemorrhage	5	1.8
None of the above	4	1.4
Transmission of SCD		
Food & drinks	1	.4
Contact with the affected person	1	.4
Blood Transfusion	10	3.6
Inheritance	260	93.2
None of the above	7	2.5

Table 3. Study Participants responses regarding SCD and Marriages

	Frequency	Percentages
Have you ever thought of pre-marital sickle cell disease testing?		
Yes	59	21.1
No	84	30.1
Not married & will do premarital testing	116	41.6
Not married & will not do premarital testing	20	7.2
If you answered Yes; is SCD testing painful and difficult?		
Strongly agree	13	9.0
Agree	14	9.7
Neutral	44	30.6
Disagree	27	18.8
Strongly disagree	46	31.9
If you recognize your fiancé/fiancée is SCD +ve, would you still marry him/her?		
Yes	25	9.0
No	176	63.5
I don't know	76	27.5
Causes of consanguineous marriage		
Marriage within the family reduce the doubts between the two partners	61	22.9
Strengthen family relationships	107	40.3
Lessens the financial burdens of marriage	25	9.4
Similarity in lifestyles	73	27.4
Do you think consanguineous marriages lead to		
1-Children with congenital malformation	14	5.0
2-Children with genetic blood disease	74	26.6
Both (1 and 2)	180	64.7
No risks associated	10	3.6

Students were asked if they ever thought of pre-marital sickle cell disease testing. The most frequent response was not married and will do premarital testing (41.6%) and least (7.2%) was not married and will not do premarital testing. Majority of study participants (91%) said if they will know their future spouse have SCD will not or will think about marry him or her. More than half (64.7%) think agreed that consanguineous marriages lead to children with genetic blood disease or congenital malformation.

In the questionnaire, students were asked about their perception regarding SCD (Table 4). Agreement was highest (95.5%) for the statements suggesting that it is useful to know if you have sickle cell trait and it is useful to know whether your husband/wife has sickle cell trait (96.8%). More than a half (62.9%) of student ere not agreed to pay for sickle cell disease testing. However, 77.7% students believed that if people knew about sickle cell disease complications, it will change their consanguinity practice.

Table 4. Student's Perception regarding Sickle Cell Disease (n-279)

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
Sickle Cell disease is a chronic disease	156 (56.5)	100 (36.2)	14 (5.1)	4 (1.4)	2 (0.7)
Pre-marital test must be mandatory	189 (68.5)	62 (22.5)	23 (8.3)	1 (0.4)	1 (0.4)
Benefits					
It is useful to know if you have sickle cell trait	197 (71.1)	68 (24.4)	11 (3.9)	1 (0.4)	0
It is useful to know whether your husband/wife has sickle cell trait	207 (74.5)	62 (22.3)	7 (2.5)	2 (0.7)	0
If you know the risk of your child having sickle cell disease, will this affect your future pregnancy plans?	132 (47.5)	85 (30.5)	53 (19)	6 (2.2)	2 (0.7)
Barriers					
It is hard to convince my partner to get tested for sickle cell disease	32 (11.6)	66 (23.8)	88 (31.5)	50 (17.9)	41 (14.7)
I would not pay for sickle cell disease testing	28 (10)	27 (9.7)	47 (17)	81 (29.2)	94 (33.7)
Consanguinity & sickle cell disease					
Do you support marrying relatives?	12 (4.4)	29 (10.7)	118 (43.4)	63 (22.6)	50 (17.9)
Do you think there is a relationship between sickle cell disease and consanguinity?	115 (42)	109 (39.8)	44 (16.1)	4 (1.5)	2 (0.7)
I would not be bothered to get tested for sickle cell disease if my husband/wife was not a relative	28 (10.1)	31 (11.2)	55 (19.9)	82 (29.6)	81 (29.2)
If people knew about sickle cell disease complications, it will change their consanguinity habits?	114 (41)	102 (36.7)	45 (16.2)	14 (5)	3 (1.1)

DISCUSSION

Sickle cell disease is a group of genetic diseases prevalent in tropical and subtropical regions as achronic haemolytic disorder caused by homozygous inheritance of abnormal haemoglobin called 'haemoglobin S' (HbS). The disease burden is enormous to the patient, family and community (Modell, 2008). Majority of participants in this study are female, only 7.5% married, 29.1% tested for SCD while rest of the participants did not have screening for SCD. General knowledge and perception regarding SCD is adequate in medical and pharmacy students however, the result shows specific knowledge about disease is not sufficient. Total prevalence of SCD is around 20% and 25% chance to have effect in children if parents have SCD.As reported in the literature the same findings regarding positive attitude towards SCD and had the belief that it is an inherited disease acquired from parents but, there was poor understanding and inadequate knowledge of SCD particularly on the pattern of inheritance. Literature has reported that doctors have a statistically significantly better knowledge of best time for detecting genotype than others (Boadu, 2018 and Animasahun, 2009).

Most of our study participants have correct and appropriate knowledge regarding pre-marital carrier screening (PMCS), however only29.1% had screening done showing 3.8% of SCD and 22.8% SC trait. Around one third of the participants were unaware of PMCS. More than one third participants were not married but willing to do PMCS. In this study the students urged not to marry a person with SCD or SCT.Taif University students have shown positive attitude and good intended practices toward PMS.Saudi government has implemented a mandatory premarital screening (PMS) program. Targeted educational programs about the importance of PMCS are strongly recommended. In high prevalent areas of SCD, the awareness should start in school (Melaibari, 2017). Oludari from Nigeriareported that significant association between respondents' educational qualification and knowledge, attitude and practices related to SCD and SCD premarital counseling, and between age and attitude and practices related to SCD premarital counseling (Oludarei, 2013 Emmanuel , 2009 and Ezechukwu, 2004). Though most of the participants were aware of premarital genotype screening and geneticcounseling, some of them lack adequatecomprehensive knowledge about it (Ugwu, 2016).

One study from Oman reported positive attitude towards PMS but inadequate knowledge about which diseases it targets. Even though the vast majority of them thought it is important to carry out PMS and agreed to do it (Al Kindi, 2012). In our study nearly two third participants willing not to marry a person with SCD as this increases the chance to have children with SCD, however the practice is not appropriate as it is very difficult to convince the partner for PMCS. Consanguineous marriage is very common in Oman, most of the participants of our study feel that it is good and convenient in terms of family relationship, reduces the financial burden of marriage and give a good life style. Nearly half of our study subjects do not support consanguineous marriage. Consanguinity is still the most common risk factor for a high frequency ofautosomal recessive diseases and congenital malformations (Bittles, 2010). One study from Oman reported that although the population is aware of consanguineous marriage related problem but still the marriage rate is around 49% (Mazharul Islam, 2013 and Hamamy, 2003).

Literature has reported that sickle cell disease in Oman has shown frequent complications like vaso-occlusive crisis requiring hospitalization ,acute chest syndrome, avascular necrosis of bone, splenic sequestration and dactylitis (Gihan Adly Rajappa, 2008). The chronic complications included sickle hepatopathy and sickle nephropathy, leg ulcer and priapism. SCDis one of the most common hereditary haemoglobinopathy with most cases from African and or Arab-Indian origin. Other regions also reported same complications (Wali, 2017; El-Hazmi, 2006 and Al-Gazali, 2006). Medical and pharmacystudents are better exposed to issues concerning the prevention andcontrol of sickle cell disease and other healthissues compared to students from otherfaculties. To reduce the incidence of SCD, effective public health education should be carried out places such as schools, colleges,media (radio/Television) and newspapers.

Conclusion

This study showed adequate knowledge of SCD among the studied subjects; however the attitude and practices is yet to be changed. Premarital counseling is well placed in Oman health system in primary care health checkups. Prevention of the disease through carrier identification and genetic counselling remains the only realistic approach to reduce the impact of the disease. There is a need for more emphasis on health education

and awareness program promoting sickle cell education and screening. Awareness regarding consanguineous marriage outcome must be communicated to larger population and the provision of genetic counselling to all SCD patients and carriers should be reemphasize for the identification and care of the couples at risk.

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