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Perceptions about Sickle Cell Disease among Adults in Albaha Region: A Cross-sectional Study

Abdulrahman A.M. Alghamdi, Ali M.A. Alamri, Abdulrahman H.A. Alghamdi, Saeed Y.S. Alghamdi, Fares A.M. Alzahrani, Saeed A.S. Alzahrani, Ayedh Mobarak Albishi
Faculty of Medicine, Albaha University, Albaha, Saudi Arabia

ABSTRACT

Background: Sickle-cell disease (SCD) is a group of genetic disorders characterized by abnormally shaped red blood cells (RBCs), which are destroyed at increased rates, leading to anemia. Recently, Saudi Arabia has been reported to have an increased prevalence of SCD.

Objective: The aim of the present study was to assess perceptions about SCD among the general population in Albaha region, Saudi Arabia, and the possible relationship between their levels of knowledge and socio-demographic characteristics. **Methods:** The study was carried out on 218 Saudi subjects who completed the self-administered questionnaires during the study period. **Results:** This study demonstrated that more than two-thirds of the respondents (68.80%) had good knowledge about basics of sickle cell disease. Those with poor knowledge (31.20%) were mainly younger males and works as businessmen and housewives, and all of them had no previous experience with SCD child. Despite the recorded favorable level of knowledge, certain gaps in knowledge were identified; most of them falsely believed that some foods like fava beans, lentils, falafel made with fava beans, vigna and /or nuts could precipitate sickle-cell crisis.Moreover, in depth information of our study population about pattern of inheritance of SCD and its complications were not sufficient.

Conclusion: In our study, a good level of awareness regarding SCD was found. Some of the respondents were confused about the nature of inheritance, complications and dietary factors related to the disease. So, community health education meetings and/or media programs to increase the public awareness are recommended.

Keywords: sickle-cell disease; awareness; perception; misconceptions; survey; Albaha; Saudi Arabia.

INTRODUCTION

Sickle-cell disease (SCD) is a group of genetic disorders characterized by abnormally shaped red blood cells (RBCs), which are removed from the circulation and destroyed at increased rates, leading to anemia[(1)]. The presence of hemoglobin S is the main pathological factor for the development of SCD. When hemoglobin S becomes deoxygenated, it forms aggregates with other hemoglobin molecules within the RBCs[(2)]. In addition to anemia, SCD can lead to blood vessels obstruction and infarction of different body organs. A point mutation in the gene coding the β chain of the hemoglobin molecule resulted in a single amino acid substitution (valine for glutamic acid), which leads to hemoglobin S. It is estimated that more than 200 000 babies are born with the disease each year in Africa alone[(3)]. In addition, it is a prevalent disorder among those from Mediterranean area countries like Turkey and the Arabian Peninsula[(4)].

Recently, a marked improvement in life expectancy and quality of life among SCD patients have been reported, which is mainly attributed to the advances in general prophylactic and other corrective measures[(5)]. However, proper public health awareness about SCD and its complications remains an important item in the management of SCD. Early community-based surveys conducted on African Americans in large urban areas demonstrated limited awareness of SCD in these communities[(6)]. In addition, **Adewoyin and colleagues**[(7)] demonstrated a moderate level of public health knowledge regarding SCD in Nigeria.

In the Middle East, **Al Arrayed & Al Hajeri**[(8)] reported a good level of knowledge about SCD among the public in Bahrain, and there are wide acceptance and appreciation of the SCD prevention campaigns being conducted in Bahrain.

Recently, Saudi Arabia has been reported to have an increased prevalence of SCD. The carrier status for SCD ranged from 2% to 27%, and up to 1.4% had SCD in Saudi Arabia[(9)]. In addition, the prevalence of consanguinity ranges from about 60 percent in Saudi Arabia up to 90 percent in some Bedouin communities[(10)]. Therefore, it is important to assess the perception of the public in Saudi Arabia about the SCD.

METHODS

Ethical considerations

This study was conducted after approval of the Institutional Review Board of the Faculty of Medicine, Albaha University. All participants

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received a detailed explanation that included the study purpose and methods. A written informed consent was taken from each participant. The study conserved participants' privacy, and confidentiality of the data was maintained by making code numbers for each participant.

Study design

The current study had a cross-sectional design, and it was carried out among adults' residents in Albaha Region of the Kingdom of Saudi Arabia. Medical or paramedical personnel and non-Saudi subjects were excluded from the study. The study was conducted during the period from July, 2017 to September, 2017 to assess awareness regarding SCD among males and femalesSaudi adults.

Data collection instrument

We used a self-administrated questionnaire that was adapted from previously published studies[11,12]. The questionnaire consisted of two parts: (1) Socio-demographic information and previous experience with SCD and (2) Knowledge about SCD and its complications, which was measured by closed-ended questions.

Statistical analysis:

The data were analyzed using statistical package for social sciences (SPSS) software version 20. Qualitative variables were presented as numbers and percentages in brackets,whereas quantitative variables were represented as median and interquartile range. Chi-square test was used to determine the association between two variables. A score of one was given for each correct answer and zero for wrong or don't know answers then thetotalknowledge score for each participantwas then calculated. Any score above the mean was considered as good knowledge while a score below the mean was considered poor knowledge.p value <.05 was considered significant.

RESULTS

This study was carried out on 218 Saudi adults residing in Albaha region who completed the self-administered questionnaires during the study period. There were 134 (61.5%) males and 84 (38.5%) females. Their ages ranged from 16 to 64 years with a median age of 33.00. Most of them (32.7%) were professionals; including surgeons, nurses, teachers, policemen and others), students (27.7%) and retired persons (15.8%) as shown in **table 1**.

Table 1:Socio-demographic characteristics of the studied subjects.

		N	%
Sex	Female	84	38.5%
	Male	134	61.5%
Occupation	Businessman	24	11.9%
	Housewife	24	11.9%
	professional	66	32.7%
	Retired	32	15.8%
	Student	56	27.7%
Age (years)	Range	16.00-64.00	
	Median	33.00	
	IQR	26.00-47.00	

Table 2 shows participant's perceptions about SCD, most of the study population (83.5%) had heard about SCD, mainly from the internet and their relatives (27.3% and 24.2% respectively). High percentages of (86.8%) the respondents knew that SCD is a blood disease, and most of them (66.0%) did not know if there are racial differences in the incidence of this disorder. There was good information about the cause of SCD; 198 (90.8%) knew that SCD is a hereditary disorder, and about two-third of them (68.6%) attributed it to marriage of two persons having the trait of sickle cell hemoglobin. Moreover, 39.8% recognized that SCD sometimes skip generations in families.

Participants who had previous experience with SCD child showed significantly higher level of knowledge (p<.05), and their source of knowledge was mainly primary health care (58.3%) followed by the hospital (16.7%). Nonetheless, most of the study participants did not have in depth information of SCD genetics. They had no clear knowledge about c or beta thalassemia traits. Most of the enrolled subjects regarded fever, infection and high altitudes as precipitating factors of sickle cell crisis. But, misconceptions about some foods like fava beans, lentils, falafel made with fava beans, vigna and /or nuts as precipitating factors of the crisis were highly frequent among the study participants. Unexpectedly, these false beliefs were significantly higher among subjects with previous experience with SCD child compared to their counterparts as demonstrated in **table 3**.

Table 2: Perceptions about sickle cell disease.

		Previous experience with SCD child?						
		Yes N=48		No N=170		Total N= 218		
		N	%	N	%	N	%	P value
Have you ever heard of SCD?	Yes	48	100.0%	134	78.8%	182	83.5%	<.001*
	No	0	0.0%	36	21.2%	36	16.5%	
Sources of information about SCD?	Relatives	6	12.5%	42	28.0%	48	24.2%	<.001*
	Primary health care	28	58.3%	0	0.0%	28	14.1%	
	Magazines	0	0.0%	12	8.0%	12	6.1%	
	Internet	6	12.5%	48	32.0%	54	27.3%	
	Hospital	8	16.7%	12	8.0%	20	10.1%	
	Friends	0	0.0%	24	16.0%	24	12.1%	
	Brochure	0	0.0%	6	4.0%	6	3.0%	
	Others	0	0.0%	6	4.0%	6	3.0%	
Which of the following are true of SCD?	SCD is a blood disease.	48	100.0%	136	82.9%	184	86.8%	.004*
	SCD can be identified by a blood test.	0	0.0%	20	12.2%	20	9.4%	
	Blood transfusions are an important way of treating SCD	0	0.0%	8	4.9%	8	3.8%	
Who gets SCD?	Mostly Africans	14	29.2%	44	26.8%	58	27.4%	.111
	All races are equally as likely	0	0.0%	14	8.5%	14	6.6%	
	Don't Know/ refused	34	70.8%	106	64.6%	140	66.0%	
How do you get SCD?	You are born with it (It's hereditary)	48	100.0%	150	88.2%	198	90.8%	.009*
	Don't know/refused	0	0.0%	20	11.8%	20	9.2%	
A baby will be born with SCD when	healthy person is married to someone with the trait	0	0.0%	14	9.0%	14	6.9%	<.001*
	Person with the trait married another with the trait	48	100.0%	92	59.0%	140	68.6%	
	Person with disease married to someone with the trait	0	0.0%	12	7.7%	12	5.9%	
	Healthy person married to someone with the disease	0	0.0%	38	24.4%	38	18.6%	
Does SCD sometimes skip generations in families?	Yes	40	83.3%	34	24.6%	74	39.8%	<.001*
	No		0.0%	6	4.3%	6	3.2%	
	Maybe		16.7%	98	71.0%	106	57.0%	

Table 3:Information of the studied subjects about heredity and precipitating factors of sickle cell disease.

		Previous experience with SCD child?						
		Yes N=48		No N=170		Total N= 218		
		N	%	N	%	N	%	P value
Do you know if there are different types of traits that can lead to SCD?	Yes	10	20.8%	18	13.0%	28	15.1%	.003*
	No	28	58.3%	52	37.7%	80	43.0%	
	Maybe	10	20.8%	68	49.3%	78	41.9%	
Do you know, if you, personally, have sickle cell trait?	Yes	42	87.5%	0	0.0%	42	20.6%	<.001*
	No	6	12.5%	100	64.1%	106	52.0%	
	Maybe	0	0.0%	56	35.9%	56	27.5%	
Have you ever heard of C-trait?	Yes	0	0.0%	14	8.5%	14	6.6%	.043*
	No	48	100.0%	150	91.5%	198	93.4%	
Do you know, if you, personally, have C-trait?	No	6	27.3%	26	32.5%	32	31.4%	.640
	I do not know	16	72.7%	54	67.5%	70	68.6%	
Have you ever heard of b-thalassemia trait?	Yes	48	100.0%	104	63.4%	152	71.7%	<.001*
	No	0	0.0%	60	36.6%	60	28.3%	
Precipitating factors of sickle cell crisis include	Traveling at high altitudes	0	0.0%	12	8.0%	12	6.1%	.002*
	Infection	0	0.0%	8	5.3%	8	4.0%	
	Fever, Infection	0	0.0%	14	9.3%	14	7.1%	
	combination	48	100.0%	116	77.3%	164	82.8%	
Patients with SCD should abstain from the following foods to avoid a crisis	fava beans	6	15.0%	6	8.3%	12	10.7%	<.001*
	Nuts	0	0.0%	6	8.3%	6	5.4%	
	Nuts, Peanuts	0	0.0%	8	11.1%	8	7.1%	
	Lentils	0	0.0%	14	19.4%	14	12.5%	
	Kidney beans (Phaseolus)	0	0.0%	6	8.3%	6	5.4%	
	Falafel made with fava beans	0	0.0%	6	8.3%	6	5.4%	
	Vigna (Lobia), Nuts, Peanuts	0	0.0%	6	8.3%	6	5.4%	
	combination	34	85.0%	20	27.8%	54	48.2%	

Knowledge of the study participants about complications of SCD was demonstrated in **table 4**, most of the study population (74.5%) identified that SCD causes severe pain that requires hospitalization. But, great percentages (72.7%, 75.8%, 85.9% and 54.9%) did not know accurately that SCD could decrease the child school performance, lead to stroke, renal failure or life threatening infections. Though, participants with previous experience with SCD child showed significantly ($p<.05$) higher levels of information about SCD complications. Moreover, all of them (100%) stated that there is no currently a cure for SCD. Finally, most of the surveyed subjects suggested health education of the public through meetings, TV and /or distribution of videos or CDs as methods to increase awareness about SCD. Total knowledge score was calculated, it ranged from 3 to 27 with a median of 22.00, (IQR: 21.00- 24.00). **Table 5** shows the association between the levels of knowledge of the study population with their socio-demographic characteristics. Participants with good knowledge had non-significantly higher median age ($P >0.05$), 56.0% of whom were males ($P < 0.05$), and 71.6% were professionals and students ($P < 0.05$). On the other hand, of those with poor knowledge 73.5% were males, 35.3% were businessmen, and all of them (100%) had no previous experience with SCD child.

Table 4: Knowledge of the study participants about complications of sickle cell disease.

		Previous experience with SCD child?						
		Yes N=48		No N=170		Total N= 218		
		N	%	N	%	N	%	P value
Does pain in SCD require hospitalization?	Yes	48	100.0%	104	66.7%	152	74.5%	<.001*
	Maybe	0	0.0%	52	33.3%	52	25.5%	
Does SCD leads to Poor school performance?	Yes	14	29.2%	40	26.7%	54	27.3%	.735
	Maybe	34	70.8%	110	73.3%	144	72.7%	
Does SCD lead to Stroke?	Yes	22	45.8%	26	17.3%	48	24.2%	<.001*
	Maybe	26	54.2%	124	82.7%	150	75.8%	
Does SCD lead to Kidney failure?	Yes	16	33.3%	12	8.0%	28	14.1%	<.001*
	Maybe	32	66.7%	138	92.0%	170	85.9%	
Does SCD lead to Life threatening infections?	Yes	36	75.0%	44	28.2%	80	39.2%	<.001*
	No	6	12.5%	6	3.8%	12	5.9%	
	Maybe	6	12.5%	106	67.9%	112	54.9%	
Is there currently a cure for SCD?	Yes	0	0.0%	16	10.7%	16	8.1%	.014*
	No	48	100.0%	134	89.3%	182	91.9%	
the best way to increase awareness about SCD in the community	Publicize on radio	0	0.0%	12	7.1%	12	5.5%	.043*
	Mail out written information or pamphlet	0	0.0%	6	3.5%	6	2.8%	
	Hold informational meetings in community	16	33.3%	52	30.6%	68	31.2%	
	Distribute a video or CD about SCD	18	37.5%	50	29.4%	68	31.2%	
	Don't know/refused	0	0.0%	12	7.1%	12	5.5%	
	Other	0	0.0%	6	3.5%	6	2.8%	
	Publicize on TV	14	29.2%	32	18.8%	46	21.1%	

Table 5: Associations between levels of knowledge among the studied subjects with their Socio-demographic characteristics.

			Level of knowledge		
			Good N=150 (68.80%)	poor N=68 (31.20%)	P value
Age	Minimum		16.00-64.00	24.00-63.00	.871
	Median		35.00	31.00	
	IQR		26.00-47.00	27.00-47.00	
	Mean rank		109.03	110.53	
Sex	Female	N	66	18	.014*
		%	44.0%	26.5%	
	Male	N	84	50	
		%	56.0%	73.5%	
Occupation	Businessman	N	0	24	<.001*
		%	0.0%	35.3%	
	Housewife	N	12	12	
		%	9.0%	17.6%	
	professional	N	48	18	
		%	35.8%	26.5%	
	Retired	N	26	6	
		%	19.4%	8.8%	
	Student	N	48	8	
		%	35.8%	11.8%	
Experience with SCD child	Yes	N	48	0	<.001*
		%	32.0%	0.0%	
		N	102	68	
		%	68.0%	100.0%	

DISCUSSION

This study demonstrated that more than two-thirds of the respondents (68.80%) had good knowledge about basics of sickle cell disease. In consistence with this, a study in Bahrain carried by **Al Arrayed and Al Hajeri**[8],showed good level of knowledge about SCD among the public. In addition, **Treadwell *et al*.**[13] reported that 68% of their study population responded correctly to knowledge questions about SCD. In contrast, lower level of knowledge was reported among SCD patients in Al-Qatif area, Eastern Province, Saudi Arabia and secondary school students in Nigeria[12,14].

Those with poor knowledge (31.20%) were non-significantly younger, mainly males and works as businessmen and housewives. This finding agrees with that of **Al Arrayed and Al Hajeri**[8]**,** who found that females gave significantly more correct answers than males. Additionally, all of them had no previous experience with SCD child.

Similarfinding was reported by **Acharya *et al*.**[15] among American parents. Based on these findings, educational campaigns targeting males in different occupations are required.

Despite this favorable level of knowledge among Saudi general population in Albaha region, there were certain misconceptions about SCD.

Most of them falsely believed that some foods like fava beans, lentils, falafel made with fava beans, vigna and /or nuts could precipitate sickle-cell crisis. Unexpectedly, these false beliefs were significantly higher among subjects with previous experience with SCD child compared to their counterparts. Similarly, **Al-Suwaid*et al*.**[12]and**Al Arrayed and Al Hajeri**[8] showed that almost two-thirds of their study participants had the same false belief. However, in fact, food items do not trigger SCD crises unless there is accompanying G-6-PD deficiency.

In which case, only the consumption of fava beans would trigger an attack[8]. Moreover, in depth

information of our study population about pattern of inheritance of SCD and its complications were not sufficient. These findings should be targeted in the health education programs.

The source of information of the study population was mainly the internet (27.3%) and relatives (24.2%) however, those who has SCD child gained theirknowledge mainly from primary health care (58.3%) followed by the hospital (16.7%). This should be considered throughout preparation of intervention programs to increase the public awareness about SCD.

CONCLUSION

In conclusion, a good level of awareness regarding SCD in our study sample was found. Some of the respondents were confused about the difference between the carrierstate of a disease and the affected state. Most of the respondents support and appreciate community health education meetings and/or TV programs to increase the public awareness. Informational programs should target the male population, and emphasizeon the nature of inheritance of the common blood diseases, its complications and dietary factors related to the disease.

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