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Knowledge and Awareness of Sickle Cell Anemia: Cross-Sectional Study among the General Population in Saudi Arabia

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ABSTRACT

Sickle cell disease (SCD) is a hereditary blood disorder. Consanguineous marriage gave a high incidence of the disease. **Objective:** the current study was to build interest to Knowledge assessment & awareness of Sickle Cell Anemia (SCA) in different regions in Saudi society and try to spread more information about it. Also, assessing the willingness of society to do premarital testing to prevent genetic diseases. **Methods:** An electronic questionnaire was circulated on social media to male and female members of Saudi Arabia's population from different age groups. A Cross-sectional study was conducted from October 2021 to December 2021. A total number of 400 participants filled out the questionnaire. Utilizing existing literature, practicing physicians, & hematologists. **Results:** Number of participants in the 20-30 age group; Regarding gender, 104 (26%) males and 296 (74%) females. For marital status ,259 (64.7%). unmarried, 133 (33.3%) married. 4 (1%), divorced & 4 (1%) widowed. For the Educational level, a university level of education; 307 (76.8%), 67 (16.7%) high school level. 16 (4%) advanced level. 6 (1.5%) middle school level. 4 (1%) elementary level. Regarding place of residence, from Hail region; 234 (58.5%) then other regions; 62 (15.5%), Riyadh region; 39 (9.8%), Almadinah Almonawara region;27 (6.7%), Alshargyah region; 22 (5.5%) and 4% from Makkah region. **Conclusions:** The overall percentage of awareness was 60.16%. Only 53 (13.3%) recognized the effects of disease on children in case both parents were affected. Most agreed about the necessity of pre-marital examination, 390 (97.5%).

INTRODUCTION

Sickle cell disease is a hematological disorder affecting people around the world. In SCD, substitution in the β-globin chain polymerizes mutant hemoglobin S, impairing erythrocyte survival and rheology. Clinically, hemolytic anemia and microvascular vaso-occlusion cycles lead to end-organ ischemia-reperfusion injury and infarction causing vasculopathy due to inflammation and redox instability [1-2] Parenchymal injury and chronic organ damage follow repeated sickling & continuing hemolytic anemia, resulting in substantial morbidity & mortality [3]. A constant state of vaso-occlusion and inflammation & the resulting progressive damage to many organs, become apparent with increasing age. The earliest sign of SCD is often dactylitis [4-5]. Complications were seen in SCD patients such as Vaso-occlusive crisis, Acute chest syndrome & Acute splenic sequestration. Patients received crizanlizumab therapy and experienced fewer pain crises concerning sickle cell disease than placebo, as well as fewer adverse events [6]. Chest x-rays showing a nary infiltrate with or without pain, cough, fever, or hypoxia are signs of ACS. Associated with acute anemia is acute splenic sequestration. [7- 8]. Early diagnosis & preventive measures can help patients with sickle cell disease, like prophylactic penicillin, blood transfusions, and hematopoietic stem-cell transplantation. The better prediction of SCD severity, the more precise treatment, & management for the patient [9-10]. SCD comes in different forms, the most common & severe form is HbSS. Hb S beta-thalassemia is very rare. [11].SCD is an inherited autosomal co-dominant trait; individuals who have the SCT (HbAS) in the heterozygous form are not affected by SCD, whereas those who carry the βS allele are affected by SCD [8]. Globally, the prevalence of sickle cell disease is 111.91 per 100,000 lives. In Saudi, among adults, it is estimated that

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(4.2%) have SCT and (0.26%) have SCD, with the eastern region having the highest prevalence. Consanguineous marriage gave a high incidence of the disease. The pre-marital program improved population awareness and knowledge of SCD, every region of Saudi Arabia has seen a decrease in SCD. Despite this, the prevalence remains higher than in other countries [12,13].

METHODS

An electronic questionnaire was circulated on social media to male and female members of Saudi Arabia's population from different age groups. A Cross-sectional study was conducted from October 2021 to December 2021. A total number of 400 participants filled out the questionnaire. Utilizing existing literature, practicing physicians, & hematologists. An informed consent form and 19 questions assessed participants' knowledge and awareness of SCD, as well as society's willingness to perform premarital testing to prevent genetic disease.

RESULTS

In total, 400 participants answered the questionnaire. Table I Showed demographic characteristics among the studied population, KSA, 2021. Number of participants in the 20-30 age group; 214 (53.5%). & less than 20 years; 74 (18.5%). The 30-40 years group; 53 (13.2%). The 40-50 years group; 33(8.3%). & More than 50 years;26 (6.5%). Regarding gender, 104 (26%) males and 296 (74%) females. For marital status ,259 (64.7%). unmarried, 133 (33.3%) married. 4 (1%), divorced & 4 (1%) widowed. For the Educational level, a university level of education; 307 (76.8%), 67 (16.7%) high school level. 16 (4%) advanced level. 6 (1.5%) middle school level. 4 (1%) elementary level. Regarding place of residence, from Hail region; 234 (58.5%) then other regions; 62 (15.5%), Riyadh region; 39 (9.8%), Almadinah Almonawara region;27 (6.7%), Alshargyah region; 22 (5.5%) and 4% from Makkah region.

	Frequency (No.)	Percent (%)
Age group		
20>	74	18.5
20- 30	214	53.5
30- 40	53	13.2
40- 50	33	8.3
50<	26	6.5
Gender		
Male	104	26.0
Female	296	74.0
Place of residence		
Hail region	234	58.5
Riyadh region	39	9.8
Alshargyah region	22	5.5
Almadinah region	27	6.7
Makkah region	16	4.0
Other regions	62	15.5
Marital status		
Single	259	64.7
Married	133	33.3
Divorced	4	1.0
Widowed	4	1.0
Educational level		
Illiterate	0	0
Elementary school	4	1.0
Middle school	6	1.5
High school	67	16.7
University	307	76.8
Advanced studies	16	4.0

Table 1: Demographic characteristics among the studied population, KSA, 2021(n=400)

Table 2 Shows the frequency and rate of responses. Most respondents (75%) said YES when asked about hearing of SCD before, most were aware of SCD classification as "blood disease" 343 (85.8%), 23 (5.8%) answered other, 13 (3.2%) bone disease, 10 (2.5%) lymphatic disease, 6 (1.5%) nerve disease, & 5 (1.2%) gastrointestinal disease. When asked if they had SCD, most answered NO; 396 (99%) & 4 (1%) YES. When asked if they knew anyone who had SCD, most of them answered NO; 379 (94.8%), only 21 (5.2%) answered YES. 6 of them (24%), and 6 (24%) were their cousins/friends respectively, 5 (20%) were their brother/sister, 4 (16%); were other, 3 (12%) were aunt/uncle, & only 1 (4%) were mother/father. When asked about how people got affected, 210 (52.5%) answers "inherited". When asked about the severity of SCD, 156 (39%) answered moderate, 148 (37%) very serious, 82 (20.5%) didn't know, & 14 (3.5%) denied its seriousness. Only, 95 (23.8%) knew that SCD is incurable. 302 (75.5%) answered that blood tests were the diagnostic method of SCD. However, only 53 (13.3%) recognized the effects of disease on children in case both parents were affected. Most agreed about the necessity of pre-marital examination, 390 (97.5%) When asked if participants' decision of marriage would be affected by their partner's results, most; 344 (86%) answered YES. They were asked what they should do if their genetic test showed a chance of having a child with SCD, 213 (53.2%) chose to consult a doctor and only 148 (37%) chose separation.

Have you heard about (Sickle Cell Disease) before?		
Q\A	Number	%
Yes	300	75.0
No	100	25.0
What kind of diseases is SCD classified?		
Bone disease	13	3.2

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Gastrointestinal disease	5	1.2
Blood disease (correct answer)	34	85.8
Lymphatic diseaseNerve disease	310	2.5
Nerve disease	6	1.5
Others	23	5.8
Yes	**Do you have SCD?**	
No	43	1.0
	96	99.0
	Do you know someone who has SCD?	
Yes	21	5.2
No	379	94.8
	If yes? how do you know them	
Mother/Father	1	4.0
Brother/Sister	5	20.0
Aunt/Uncle	3	12.0
cousin	6	24.0
friend	6	24.0
other	4	16.0
	How do you think people with SCD get affected?	
Malnutrition	37	9.2
By virus	16	4.0
By bacteria	20	5.0
Inherited (correct answer)	210	52.5
Mineral deficiency	26	6.5
Other	3	0.8
I do not know	88	22.0
	What do you think about the severity of SCD?	
Very serious	148	37.0
moderate	156	39.0
Not serious	14	3.5
I do not know	82	20.5
	Is SCD curable?	
Yes	96	24.0
No (correct answer)	95	23.8
I do not know	209	52.2
	Which one of those is the test to diagnose SCD?	
Ultrasound	9	2.2
Blood test (correct answer)	302	75.5
Urine / feces test	7	1.8
Others	0	0.0
I do not know	82	20.5
	What is the probability for a child to get SCD? if both parents had SC trait	
All children	83	20.8
50% of offspring	109	27.2
(Correct answer)25% of offspring	53	13.3
I do not know	155	38.7
	Do you think pre-marital examination is necessary?	
Yes	390	97.5
No	10	2.5
	Does your partner result affect your marriage decision?	
Yes	344	86.0
No	56	14.0
	If a married couple found out that their genetic test showed the chance of having a child with SCD ,what do you think they should do	
separate	148	37.0
Continue their marriage	7	1.8
Consult a doctor (correct answer)	213	53.2
I do not know	32	8.0

Table 2: Participants' answers and percentages for SCD Awareness questions (n=400)

Q\A	Age					P-value
	20>	20-30	30-40	40-50	50<	
Have you heard about (SCD) before						
Yes	51	165	38	25	21	0.602
No	23	49	15	8	5	
What kind of diseases is SCD classified						
Bone	6	6	0	1	0	0.096
GI	3	1	1	0	0	
Blood	53	190	47	29	24	
lymphatic	4	4	0	1	1	
Nerve	1	5	0	0	0	
Others	7	8	5	2	1	
How do you think people with SCD get affected						
malnutrition	4	15	10	4	4	0.013
By virus	2	11	3	0	0	
By bacteria	7	10	3	0	0	
Inherited	35	117	21	22	15	
Mineral deficiency	5	17	3	0	1	
others	0	1	0	2	0	
I do not know	21	43	13	5	6	
What do you think the severity of SCD is						
Very severe	26	68	24	18	12	0.141
Moderate	27	97	13	9	10	
Not serious	3	9	1	1	0	
I do not know	18	40	15	5	4	
Is it curable						
Yes	17	47	14	8	10	0.485
No	13	59	10	8	5	
Yes	17	47	14	8	10	0.485
No	13	59	10	8	5	
I do not know	44	108	29	17	11	
Which one of those is the test to diagnose SCD						
Ultrasound	6	3	0	0	0	0.024
Blood test	49	168	35	28	22	
Urine feces test	2	3	2	0	0	
I do not know	17	40	16	5	4	
What is the probability for a child to get SCD, if both parents have it						
All children	13	49	11	6	4	0.859
50% of offspring	24	53	14	11	7	
25% of offspring	12	29	4	3	5	
I do not know	25	83	24	13	10	
Do you think pre marital examination is necessary						
Yes	73	209	51	32	25	0.909
No	1	5	2	1	1	
Does your partner result affect your marriage						
Yes	63	191	41	30	19	0.050
No	11	23	12	3	7	
If a married couple found out that their genetic test showed the chance of having child with SCD, what do toy think they should do						

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separate	31	73	18	13	13	0.255
Continue their marriage	2	4	0	1	0	
Consult a doctor	32	122	31	19	9	
I do not know	9	15	4	0	4	

Table 3: The correlation between awareness and age groups of the studied population

Q\A	Marital status				P-value
	Single	Married	Divorced	Widow	
Have you heard about (SCD) before					
Yes	194	99	3	4	0.716
No	65	34	1	0	
What kind of diseases is SCD					
Bone	11	2	0	0	0.437
GI	4	1	0	0	
Blood	217	119	3	4	
lymphatic	8	1	1	0	
Nerve	5	1	0	0	
Others	14	9	0	0	
How do you think people with SCD get affected					
malnutrition	15	21	0	1	0.000
By virus	11	5	0	0	
By bacteria	16	4	0	0	
Inherited	132	73	3	2	
Mineral deficiency	21	5	0	0	
others	1	1	0	1	
I do not know	63	24	1	0	
What do you think the severity of SCD is					
Very severe	88	57	1	2	0.241
Moderate	103	50	1	2	
Not serious	11	2	1	0	
I do not know	57	24	1	0	
Very severe	88	57	1	2	0.241
Moderate	103	50	1	2	
Not serious	11	2	1	0	
I do not know	57	24	1	0	
Is SCD curable					
Yes	53	42	0	1	0.191
No	65	29	1	0	
I do not know	141	62	3	3	
Which one of those is the test to diagnose SCD					
Ultrasound	9	0	0	0	0.389
Blood test	188	109	2	3	
Urine\feces test	5	2	0	0	
I do not know	57	22	2	1	
What is the probability for a child to get SCD, if both parents have it					
All children	53	30	0	0	0.663
50% of offspring	69	36	3	1	
25% of offspring	34	18	0	1	
I do not know	103	49	1	2	
Do you think pre_marital examination is necessary					
Yes	253	130	3	4	0.037
No	6	3	1	0	
Does your partner result affect your marriage					
Yes	229	109	2	4	0.046
No	30	24	2	0	

Table4: correlation between awareness of SCD and marital status

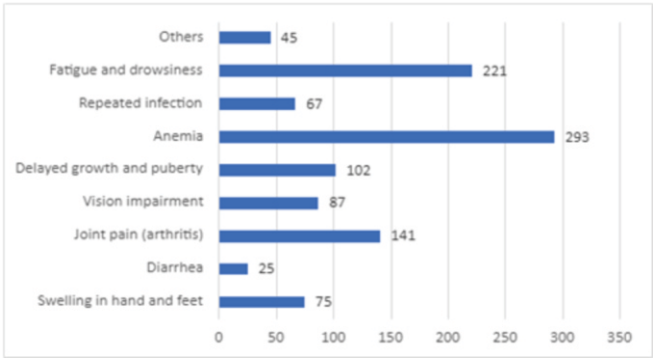

Figure 1: Demonstrated knowledge about the symptoms of SCD (n=400)

DISCUSSION

To spread more information about SCD in Saudi society, the aim of the study was to assess the knowledge of the causes, symptoms, diagnosis, & treatment of SCD in Saudi Arabia. Also, assessing the willingness of society to do premarital testing to prevent genetic diseases from spreading. Identifying the awareness campaigns needed for the targeted generations and areas, there was a variance in the contribution rate between males & females in data analysis, 74% being female. similar findings were seen in previous studies, like in Hassan, et al, 2019 [14]. This might be due to a distribution flaw where females received more requests than males, or an interested imbalance. Regarding the marital state, 64.7% were single, 33.3% were married. Divorced and widows were detected with minimum rates. Overall percentage of awareness was

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60.16%. Most of the participants had previously heard about the diseases (75%), Shaikha, Al Hajeri (2010) and Al-ghubishi, 2021 reported similar results [15,16], the percentage of awareness wasn't surprising and it's common to know people affected by SCD in KSA. As for the classification of SCD, 85.8% had answered blood disease correctly. Regarding the cause of SCD, 52.5%. Reflecting an overall suitable conception. Only 23.8% answered that SCD wasn't curable. This denoted poor knowledge about the severity of the disease. For the diagnostic methods of SCD, 75.5% answered correctly. The rest might have poor knowledge of the diagnostic measures. When asked about the probability of a child being were small percentage of 13.3% answered correctly. As for the necessity of pre-marital testing, most participants (97.5%) agreed which was consistent with Ibrahim, et al, 2011 [17]. Influence on the decision to marry them, 86% agreed. This is consistent with Maryam, et al, 2017 [18]. This can be attributed to the Saudi government's premarital screening program 53.2% declared that their susceptibility of have a child with SCD. SCD complications and difficulties facing affected families could explain this attitude. Concerning age-related awareness, the age group (40-50) followed by the age group (20-30) showed a higher level of knowledge about SCD. Participants less than (20 years) showed a poor knowledge about SCD in most questions, followed by the age group (30-40). Across all age groups, there were positive attitudes toward premarital examination. The increase a better attitude was expected with greater knowledge. Similarly, to Binshihon et al, 2018 [19]. As for the correlation of awareness with the marital state, both the single & married participants showed poor knowledge of getting affected with SCD, being curable & the probability that a child could get SCD if both parents have it. A significant difference in answer was observed when asked about the cause of SCD, p-value=0.000., & necessity of pre-marital examination, p-value = 0.037. A significant difference was noticed about the affection of marriage decision based on the partner result, p-value = 0. 046. This finding ran parallel with, Alhowiti and Shaqran, 2019 [20].

CONCLUSION

In this study Saudi Arabians of different ages in different region were assessed by Knowledge and attitude. knowledge of the disease varied from overall moderate level (66.16%) that necessitate more efforts through health education about SCD in different social media. Positive response was given toward SCD& premarital screening.

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