Medical Science

pISSN 2321–7359; eISSN 2321–7367

Community awareness of hereditary blood diseases and the benefit of premarital screening program in Saudi Arabia

Mohamed A Elhefny[1, 2], **Adnan Alessa**[3], **Saud Alzahrani**[3], **Yahya Almarhabi**[3], **Badr Almaedy**[3], **Omar Alshaikhi**[3], **Hatem Alessa**[3], **Hasna Alghamdi**[3], **Abdullah Alamri**[3], **Thamer Alghamdi**[4]

To Cite:
Elhefny MA, Alessa A, Alzahrani S, Almarhabi Y, Almaedy B, Alshaikhi O, Alessa H, Alghamdi H, Alamri A, Alghamdi T. Community awareness of hereditary blood diseases and the benefit of premarital screening program in Saudi Arabia.. Medical Science 2022; 26:ms424e2505.
doi: https://doi.org/10.54905/disssi/v26i128/ms424e2505

Authors' Affiliation:
[1]Department of Genetics, College of Medicine-AlQunfudah, UmmAl-QuraUniversity, Makkah, Saudi Arabia
[2]Department of Cancer and Molecular Biology, NCI Cairo University, Cairo, Egypt
[3]Students of College of Medicine-AlQunfudah, UmmAl-QuraUniversity, Makkah, Saudi Arabia
[4]Student of College of Medicine, Al-Baha University, Al-Baha, Saudi Arabia.

Peer-Review History
Received: 05 October 2022
Reviewed & Revised: 06/October/2022 to 18/October/2022
Accepted: 21 October 2022
Published: 24 October 2022

Peer-review Method
External peer-review was done through double-blind method.

URL: https://www.discoveryjournals.org/medicalscience

ABSTRACT

Sickle cell disease is a group of inherited syndromes characterized by the presence of hemoglobin S, abnormal hemoglobin that is poorly soluble and polymerizes when deoxygenated, which results in sickle-shaped erythrocytes. These syndromes manifest with vaso-occlusive phenomena and hemolytic anemia. SCD in Saudi Arabia, which suffers from high consanguinity marriage rates, creates a fertile environment for it to spread deep into its roots. This study has targeted the Saudi Arabian community based on wondering why after a decade and a half ago of applying to premarital screening programs still SCDs a health and social issue. By using an online questionnaire covering the kingdom's regions, surveyed all Saudi communities. A total of 11276 participants completed the questionnaire; Women represent 70.6% of the participants, while men represent 29.4%. The educational level of the participants was high with 69.8% having a university education and 27.9% having high school. The unmarried participants represent 63.8% (N = 7873), who show positive awareness and perception regarding the impact of hereditary blood diseases on quality of life and the use of the premarital examination program. Married participants represent 36.2% (N = 4091) of total participants which 44.98% (N = 1840) of them have consanguinity marriage and 55.1% (N = 2251) have no consanguinity. Remarkably, the percentage of affected children with SCD of a couple who has consanguinity marriage is almost the same percentage of affected children of couples with no consanguinity, but the severity and complications elevated more in children from consanguineous marriage.

Keywords: Sickle cell disease, hereditary blood diseases, premarital screening program, Saudi Arabia, community awareness.

1. INTRODUCTION

Hereditary hemoglobin disease (hemoglobinopathies) is a mainly autosomal recessive hereditary single gene, with the formation of structurally abnormal hemoglobin variants (sickle-cell disease) or decreased synthesis of structurally normal hemoglobin causes (β-thalassemia) (Alenazi et al., 2015). Sickle cell anemia and β-thalassemia affect up to 5% of the world's population and are a major public health concern in certain parts of the world, including the Mediterranean and the Middle East. Saudi Arabia is known for its high prevalence of hereditary blood disorders. A study published in 2007 and conducted in Saudi Arabia reported that 4.20% of participants had sickle cell trait, 0.26% had sickle cell anemia, and 3.22% had β-thalassemia trait, while 0.07% had β-thalassemia disease. In 2004, a compulsory pre-marriage genetic testing was introduced in Saudi Arabia due to the impact of the strain on the health care system on the quality of life for the patients with β-thalassemia or sickle cell disease (Memish & Saeedi, 2011). Saudi Arabia has a high prevalence (42.1–66.7%) and is considered one of the top four countries of consanguineous marriages due to several cultural factors (El-Mouzan et al., 2007).

A screening for both diseases must be done for all couples who are applying for marriage proposals and (if necessary) must be given an appropriate advice before submitting proposals. Implementation of the counseling recommendations was left voluntary. The results of this national program have not yet been quantified (Memish & Saeedi, 2011). Prenatal and neonatal screening programs for hemoglobinopathy are considered cost-effective in populations with high prevalence, particularly because of the high mortality associated with thalassemia and the financial, social, and psychological costs of maintaining patients with these diseases. However, premarital screening programs have more benefits compared to newborn screening programs which can only provide secondary or tertiary prevention for children suffering from the secongenital diseases. Prenatal screening programs can reduce morbidity but are not acceptable in some societies. Under these circumstances, premarital screening programs have great potential in reducing the incidence of genetic disorders. These diseases are not fatal but are chronic and require ongoing treatment and monitoring. Therefore, the cost is at both the national and individual levels (AlHamdan et al., 2007). Malaria endemic disease has been reported to correlate with SCD occurrence. The presence of SCD is known to provide survival benefits and protective effects against malaria.

A recently published study estimated the prevalence, 5-year time trends, and distribution of β-thalassemia and SCD in Saudi Arabia using data from the Saudi premarital screening and genetic counseling program from February 2011 to December 2015. A downward trend was observed in the prevalence of β-thalassemia across the years; however, the prevalence of SCD remained constant. One of the key goals of premarital screening and genetic counseling (PMSGC) is to reduce or eliminate the presence of β-thalassemia and SCD in newborns. One of the strengths of the Saudi PMSGC program is the fact that it is mandatory and essential for anyone who wants to get married. This will raise the awareness of the population regarding hemoglobinopathy diseases and enhances the effectiveness of this program.

The program provides extensive support, reaching 4% of the population each year, and provides valuable data resources and research opportunities. In addition to identifying changing trends in these disorders, the data contribute to a more accurate assessment of the extent of hemoglobinopathy disorders in Saudi Arabia (Alsaeed et al., 2017). The total number of marriage cancellations among risk marriages in 2004 increased from 10% to more than 50% in 2009, and 48% of risk marriages in 2009 were completed, suggesting a large cultural problem (Memish & Saeedi, 2011). A recent study evaluating the effect of sickle cell anemia on personality found depression in 48.2% of participants, and a lower educational qualification, a high incidence of vascular occlusive crises, and frequent hematology clinic visits were significantly associated with depression (Bakri et al, 2014).

Additionally, another study of 382 adults with SCD using the Beck II Depression Screening Study found that 20.6% of participants had depression, and half (10.0%) had suicidal thoughts (Wallen et al., 2014). Furthermore, other factors contribute to depression, such as low family income, multiple blood transfusions, poor pain control, inadequate social support, and a history of frequent vascular occlusive crises (Hasan et al., 2003). Effects of sickle cell anemia on the quality of life: physical health challenges of chronic SCD such as recurrent episodes of bone pain, chronic complications such as avascular necrosis negative impact on physical and general health (Nwogoh et al., 2016). A study of 285 Saudi adolescents (14-18 years of age) with SCD, who underwent routine assessment, follow-up, and management, showed that their health-related quality of life deteriorated significantly, particularly in the terms of general health and physical pain regardless of gender (Amr et al., 2011). As a result of a study of β-thalassemia cases through the pediatric quality of life test in pediatrics, it was found that it had a negative effect on the quality of life of children with thalassemia (Ayoub et al., 2013). The effect of the disease on a child impacts the family. On the other hand, family and psychological support will positively increase the child's ability to cope with the effects of chronic illness.

A study of 164 adults caring for children with SCD found that their financial and emotional burden was on those caring for these children. It affects various aspects of the quality of life that can be affected by an individual's level of social and occupational achievement (Madani et al., 2018). In general, health-related quality of life is negatively associated with frequent hospitalizations with slight variations depending on age, female gender, rural life, low family income, disease-related complications, and geographic location.

2. MATERIALS AND METHODS

A cross-sectional survey was conducted among Saudi population and was carried out by sending a modified and validated Arabic questionnaire in all Saudi Arabia regions. The questionnaires were designed in Google form and were sent online (WhatsApp or email). A cover page illustrated both the purpose of the study along with the fact that participation in the study is voluntary.

Study duration

Six months, started by July, 1st, 2021 until the end of December, 2021.

Study population

The target sample in this study was all adults, males and females, residing in Saudi Arabia.

Sample size

The sample size was calculated by Raosoft Calculator based on the size of the adult male and female population in Saudi Arabia (approximately 27, 136, and 977). To have a 95% confidence interval and a 5% margin of error, a sample size of at least 385 participants were required for this study. The questionnaire consisted of three sections. The first section focused on socioeconomic and background information such as gender, age, education level, and marital status, risk marriage with previous experience of unhealthy baby, pregnancy, and abortion. The second section was concerned mainly with the individuals' awareness and attitudes about inherited blood diseases (type, how to transmit, relation with consequently, and complications). The third section focused on an individual's awareness of the impact of a hereditary blood disease on a patient's life, disease severity and the responsibility of having a baby with a hereditary blood disease. A pilot study of randomly selected 20 adults was conducted in Al-Qunfudah city, in which the items of the questionnaire were checked thoroughly for vagueness and individuality in responses.

Data analysis

After data were extracted, revised, coded, and fed to statistical software IBM SPSS version 22 (SPSS, Inc Chicago, IL). All statistical analysis was done using two-tailed tests. A P-value less than 0.05 were statistically significant. For awareness and perception items, each composite mean was calculated for each participant that ranged from 1 to5. A participant with a composite mean score of less than three was considered to have a positive perception, while those with a composite mean score of 3 to less than 4 were in a neutral situation regarding the study issue. Descriptive analysis based on the frequency and percent distribution was done for all variables including demographic data, history of having hereditary blood diseases, consanguinity among married participants, having children with hereditary blood diseases, disease severity, complications, receiving support, and ambulance access if needed. Awareness and perception frequency distribution were assessed for all items regarding the impact of hereditary blood diseases on quality of life and the use of the pre-marital examination program to reduce the spread of genetic blood diseases. Cross tabulation was used to assess the distribution of different relations with consanguinity, having children with hereditary blood diseases, disease severity, and complications. Relations were tested using the Pearson chi-square test and the exact probability test for small frequency distributions.

3. RESULTS

A total of 11276 participants, who fulfilled the inclusion criteria, completed the study questionnaire. Nearly, 28.8% were from the Western region, 21.6% from the Central region, 19.9% from the Southern region, 15.4%b from the Western region, and 14.2% from the Northern region of Saudi Arabia. Respondents' age ranged from 18 to 67 years with a mean age of 28.6 ±12.4 years old. About 70.6% of the study participants were females, 69.8% had university-level f education or postgraduate degree. Nearly, two-thirds (62.8%) of the participants were singles, and 35% were married. As for consanguinity, 55.01% of the study group married non-relatives while 41.41% married from first cousin marriage and 3.56% married from weak consanguinity. Approximately, 4.6% were

carriers for sickle cell disease and 2.7% were diseased. In addition, 1.8% was carriers for thalassemia and 1.4% was diseased. Among participants, 30.5% had children as 15.7% had 1-3 children, 12% had 4-6 children but 2.8% had 7 children or more (table 1).

Table 1 Socio-demographic and clinical data of study participants

Bio-demographic data	No	%
Age in years		
18-29	7859	69.7%
30-39	1534	13.6%
40-49	1314	11.7%
50-59	479	4.2%
60+	90	0.8%
Gender		
Male	3312	29.4%
Female	7964	70.6%
Education level		
Below secondary	259	2.3%
Secondary	3144	27.9%
University / above	7873	69.8%
Marital status		
Single	7200	63.8%
Married	3865	34.2%
Divorced	226	2.0%
Consanguinity (n=4195)		
Not married	7200	63.8%
First cousin marriage	1702	15.1%
Weak consanguinity	146	1.3%
No consanguinity	2243	19.9%
Sickle cell disease		
Normal	10456	92.7%
Disease carrier	516	4.6%
Diseases	304	2.7%
Thalassemia		
Normal	10915	96.8%
Disease carrier	201	1.8%
Diseases	160	1.4%
Have children (n=4195)		
No	7840	69.5%
Yes	3436	30.5%
Number of children		
None	7840	69.5%
1-3	1766	15.7%
4-6	1354	12.0%
7-9	289	2.6%
10+	27	0.2%

Table 2 prevalence of hereditary diseases among children of study participants

Children's hereditary disease frequency	No	%
Have children with hereditary disease		
No	3260	94.9%
Yes	176	5.1%
Number of ill children		
None	3260	94.9%
1-3	159	4.6%
4-6	17	0.5%
The severity of the disease (n=176)		
Mild	154	87.5%
Severe	22	12.5%
The child had complications (n=176)		
No	92	52.3%
Yes	84	47.7%
Do you receive a subsidy from any government agency or charitable organization? (n=176)		
No	124	70.5%
Yes	52	29.5%
Is this subsidy sufficient? (n=52)		
No	25	48.1%
Yes	27	51.9%
Is it easy for you to access ambulance services in case you need them? (n=176)		
No	57	32.4%
Yes	119	67.6%
Do you have previous experience in abortion for unhealthy reasons for the fetus? (n=176)		
No	137	77.8%
Yes	39	22.2%

Table 2 reveals the distribution of frequency of hereditary diseases among children of study participants, in Saudi Arabia. Only 5.1% of the study participants had children with hereditary diseases, which were 1-3 children among 4.6% and 4-6 children among 0.5%. The hereditary disease was mild among 87.5% of the diseased children, and 47.7% had disease-related complications. An Exact 29.5% reported that they receive a subsidy from any government agency or charitable organization which is sufficient for 51.9%. Also, 67.6% of the participants with affected children said that it is easy for them to access ambulance services. 22.2% agreed on previous experience regarding abortion for unhealthy reasons for the fetus.

Table 3 Relation between participants' consanguinity marriage and personal data P: X^2 test * $P < 0.05$ (significant)

Personal data	Consanguinity						p-value
	First cousin marriage		Weak consanguinity		No consanguinity		
	No	%	No	%	No	%	
Age in years							0.482
18-29	1159	41.8%	104	3.8%	1510	54.5%	
30-39	240	39.7%	16	2.6%	349	57.7%	
40-49	205	41.1%	22	4.4%	272	54.5%	

50-59	80	46.8%	3	1.8%	88	51.5%	
60+	17	44.7%	1	2.6%	20	52.6%	
Education level							0.049*
Below secondary	38	39.2%	0	0.0%	59	60.8%	
Secondary	474	39.7%	53	4.4%	668	55.9%	
University / above	1189	42.6%	93	3.3%	1512	54.1%	

Table 3 demonstrates the relation between participants' consanguinity marriage and personal data. Consanguinity was reported among 50.1% of young-aged respondents compared to 38.4% of the old age group (60+) with recorded statistical significance. The exact 45.9% of highly educated married persons had consanguinity marriage compared to 53.2% of those with low education (below secondary) with no statistical significance (P=.109). Table 4 demonstrates the relation between participants' consanguinity marriage and having children with hereditary diseases. Exact 2.7% of those who had first-degree consanguinity marriage had a child with hereditary diseases and 6.8% of others with second-degree consanguinity versus 3.5% of a non-consangeous spouse with recorded statistical significance (P=.001). Disease severity and complications are insignificantly related to consanguinity marriage.

Table 4 Relation between participants' consanguinity marriage and having children with hereditary disease P: Pearson X^2 test #: Exact probability test * P < 0.05 (significant).

Child hereditary diseases	Consanguinity						p-value
	First cousin marriage		Weak consanguinity		No consanguinity		
	No	%	No	%	No	%	
Have children with hereditary disease							0.367
No	538	94.9%	37	100.0%	683	95.3%	
Yes	29	5.1%	0	0.0%	34	4.7%	
The severity of the disease							0.533#
Mild	23	79.3%	0	0.0%	29	85.3%	
Severe	6	20.7%	0	0.0%	5	14.7%	
Child had complications							0.859 #
No	13	44.8%	0	0.0%	16	47.1%	
Yes	16	55.2%	0	0.0%	18	52.9%	

Table 5 demonstrates community awareness of the impact of hereditary blood diseases on quality of life and the use of the pre-marital examination program to reduce the spread of genetic blood diseases. The highest agreement and awareness among participants was that it's vital to benefit from the PMC to avoid having children with hereditary blood diseases (96.4%). PMC is of great importance in avoiding having children with hereditary blood diseases (93.1%). Having a child with a hereditary blood disease entails greater responsibility and continuous health care (91.5%). Marriage is built on the match of the partners and the PMC compatibility to have healthy children rather than being relatives (91.4%). PMC provides assistance to those who are about to marry and helps them in making a decision (89.6%). Children with hereditary blood diseases can avoid their suffering by benefiting from PMC program (87.9%). The country and family burden increase to provide the necessary health care for children with hereditary blood diseases (83.7%). PMC mismatch indicates a high risk of children suffering from hereditary blood diseases (83.5%). On the other hand, 50.8% of the participants refused the fact that relatives' marriage is more important than the compatibility of the spouses' genes to avoid the risk of increasing genetic diseases. 38.4% refused abortion in case of fetuses with hereditary blood diseases, while 62.4% agreed that incompatible spouses should adopt contraception.

Figure 1 show overall public awareness and perception regarding the impact of hereditary blood diseases on quality of life and the use of the pre-marital examination program to reduce the spread of genetic blood diseases. The exact 6739 (59.8%) participants were aware and had a positive perception regarding the study issue of hereditary blood diseases impact and role of PMC.

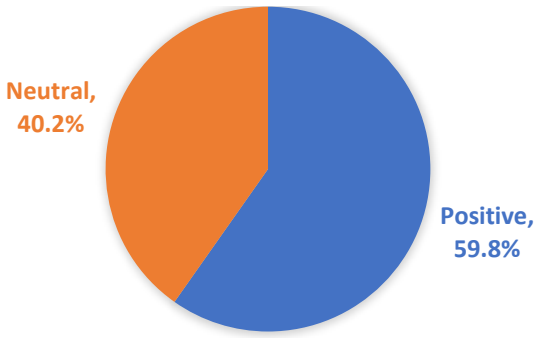

Figure 1 Overall public awareness and perception regarding impact of hereditary blood diseases on quality of life

Table 5 Community awareness of the impact of hereditary blood diseases on quality of life and the use of the pre-marital examination program to reduce the spread of genetic blood diseases

	Agree		Not sure		Disagree	
	No	%	No	%	No	%
Inconsistency of the PMC may influence my decision to proceed with the marriage	9294	82.4%	1334	11.8%	648	5.7%
It's vital to benefit from the PMC to avoid having children with hereditary blood diseases	10873	96.4%	315	2.8%	88	.8%
PMC provides assistance to those who are about to marry and helps them in making a decision	10105	89.6%	990	8.8%	181	1.6%
The interdependence of the relations of relatives by marriage is more important than the compatibility of the spouses' genes to avoid the risk of increasing genetic diseases	3732	33.1%	1819	16.1%	5725	50.8%
Marriage is built on the match of the partners and the PMC compatibility to have healthy children rather than being relatives	10302	91.4%	654	5.8%	320	2.8%
Patients with hereditary blood diseases suffer from pain with poor growth causing poor educational achievement and poor quality of life	7839	69.5%	3001	26.6%	436	3.9%
Children with hereditary blood diseases can avoid their suffering by benefiting from PMC program	9912	87.9%	1128	10.0%	236	2.1%
PMC mismatch indicates a high risk of children suffering from hereditary blood diseases	9418	83.5%	1616	14.3%	242	2.1%
PMC is of great importance in avoiding having children with hereditary blood diseases	10495	93.1%	657	5.8%	124	1.1%
Children with hereditary blood diseases have lower academic performance than children without the disease	5682	50.4%	4536	40.2%	1058	9.4%
Having a child with a hereditary blood disease entails greater responsibility and continuous health care	10322	91.5%	813	7.2%	141	1.3%
The country and family burden increases to provide the necessary health care for children with hereditary blood diseases	9440	83.7%	1464	13.0%	372	3.3%

A fetus with a hereditary blood disease necessitates its abortion to limit disease frequency and to avoid the inevitable suffering of the child	3363	29.8%	3579	31.7%	4334	38.4%
Incompatible spouses should not continue because the risk of their children suffering from a hereditary blood disease is high	7035	62.4%	2907	25.8%	1334	11.8%
Incompatible spouses should continue but should be obligated for medical counselling and fetus assessment in case of pregnancy	6567	58.2%	2359	20.9%	2350	20.8%

Table 6 Prevalence of hereditary diseases among children of the study participants

Children hereditary disease frequency	No	%
Have children with hereditary disease		
No	3260	94.9%
Yes	176	5.1%
Number of ill children		
None	3260	94.9%
1-3	159	4.6%
4-6	17	.5%
Severity of the disease (n=176)		
Mild	154	87.5%
Severe	22	12.5%
Child had complications (n=176)		
No	92	52.3%
Yes	84	47.7%
Do you receive a subsidy from any government agency or charitable organization? (n=176)		
No	124	70.5%
Yes	52	29.5%
Is this subsidy sufficient? (n=52)		
No	25	48.1%
Yes	27	51.9%
Is it easy for you to access ambulance services in case you need? (n=176)		
No	57	32.4%
Yes	119	67.6%
Do you have previous experience in abortion for unhealthy reasons for the fetus? (n=176)		
No	137	77.8%
Yes	39	22.2%

Table 6 reveals the distribution of frequency regarding hereditary diseases among children of study participants, in Saudi Arabia. Only 5.1% of the study participants had children with hereditary diseases, which were 1-3 children among 4.6% and 4-6 children among 0.5%. The hereditary disease was mild among 87.5% of the diseased children, and 47.7% had disease-related complications. An exact 29.5% reported that they receive a subsidy from any government agency or charitable organization, which is sufficient for 51.9%. Also, 67.6% of the participants with diseased children said that it is easy for them to access ambulance services in case they need them. When asked about previous experience in abortion for unhealthy reasons for the fetus, 22.2% said yes.

4. DISCUSSION

The questionnaire demonstrates good awareness of the population by prenatal program and the role for which it was created. Although 63% of responders are single, they have a high awareness of prenatal program screening because governmental procedures and requirements which do not affect the prevalence of the consanguinity marriages. Over many years prenatal screening programs and precautions has been set to minimize the percentage of inheritance diseases of at-risk marriages but the percentage hasn't changed at least as noticeable way. As our data shows that consanguinity marriage which is what we mean by at risk marriage, still the preferred way to the communities as a 41.41% are married to a First cousin, while a 3.56% are married to a weak consanguinity partner. The same result came up in previous studies, which refers that consanguinity marriages are ranging between 42.1–66.7% (El-Mouzan et al., 2007).

There is a defect in population knowledge of the extent of the effect of sickle cell disease on children. As 49.6% of responders, they do not see there is an effect on the academic performance of children with sickle cell disease. Clarify the need of need to guidance of more efforts towards educating people about the effect of sickle cell anima on the quality of life of children. There is a significant relationship between education level and non-consanguinity marriages as data analysis showed above. Clarify that educated individuals prefer non-consanguinity marriage which represents 55.01% of marriages. Many social reasons prevent a person from accepting the premarital counseling advice and annulment of his marriage and limit the usefulness of the pre-marital screening program, the most important of which is the consanguinity between the couples. Regarding the relationship between the SCA and consanguinity marriages High prevalence of SCA at 4.2% of the Saudi population (Alotaibi, 2017), which is an autosomal recessive disorder attributed to higher in consanguineous marriages this attribution can't be right (Alsaeed et al., 2017). We can't blame consanguinity as our results show (Table 6) that 4.7% of children from non-consanguinity marriages are affected with SCA compared with 5.1% of children from consanguinity marriages. Purely Mandala's genetic inheritance view will not explain the close ratio of children with SCA from both non-consanguinity marriages and consanguinity marriages. The excess risk that an autosomal recessive disorder could be expressed in the progeny of a consanguineous union is inversely proportional to the frequency of the disease allele in the total gene pool (Bittles et al., 2002).

That proved by the fact of non-consanguinity has similarly affected children to those with consanguinity demonstrates how deep the effect of consanguinity marriages which have been the tradition since the early existence of modern humans result in excess risk of Sickle cell anima trait to be expressed in population despite consanguinity absence. It is noteworthy that 10.5% of responders represent 1176 random persons from all kingdom ether carriers or has an inherence blood disease. In addition, there is a significant relationship between consanguinity marriages and more risk to have complicated severe diseases in affected children. These results were compatible with studies that discuss the Involvement of consanguinity marriages in making mutations in specific genes (e.g. Xmn1 polymorphism) that contributed to more complicated and severe SCD conditions (Kahhaleh et al., 2019; Kumkhaek et al., 2008; Wonkam et al., 2014). Although, there is a lack of this kind of study in Saudi Arabia, and more studies with larger cohorts are needed to reveal the genes that are associated with increased severity which could prepare to make a more personalized treatment (Colah et al., 2005; Theodoridou et al., 2008).

According to this study, 82.4% of the participants would cancel the marriage plan while 5.7% would continue the marriage whatever the advice was. Compared to a study conducted in 2018, the study showed that 63.5% of the participants would cancel their marriage, while 8.5% would complete it, while another study conducted in 2002 showed that 42% of the participants would complete their marriage while 27% would cancel it for a different reason. Depending on these results, there is a noticeable difference and improvement in people's awareness and acceptance of the program's advice. 96% of the participants see the benefit of the program in reducing the spread of genetic blood diseases, and according to another study, 92% believe in the effectiveness of the program in preventing the spread of genetic and infectious diseases. This is a good indicator to increase awareness and community acceptance of the program and its advice. There is also a majority prefers that the examination and counseling should be compulsory, while the choice of the decision to either cancel or complete the marriage is the right of the spouses, as it is now.

The percentage of participants who support this decision in this research is 58.2%, while the percentage in another research was 66%, On the other hand, according to a study published 20 years ago, the percentage of those who agree with this decision is 29% (Al-Khaldi et al., 2002; Binshihon et al., 2018). 62.4% of participants agreed that marriage should not be allowed between couples with a risky marriage, this opinion coincides with another research with 63.2% of participants agreeing to the same idea. Despite the ethical and social problems of making such a decision, there is a majority supporting it (Al-Aama, 2010).

5. CONCLUSION

This study shows a high incidence of consanguinity marriages despite awareness and a highly educated population. Concretely the extent of inherence to blood disease is far deep in genes of the population demonstrated by similarly affected children in both marriages, which clearly means the prenatal screening program will not be enough to limit its prevalence. For that, we suggest for more active awareness programs to inform the population about the inheritance of blood disease and its effect on the quality of life of children, which will affect their future.

Acknowledgements

The authors would like to thank the 165 participating data collectors from all over the Kingdom of Saudi Arabia.

Ethical Considerations

The study was approved by the Biomedical Research Ethics Committee, Faculty of Medicine, Umm Al-Qura University. The ethical approval code: HAPO-02-K-012-2021-03-6.

Author Contributions

The manuscript's preparation and editing were a collaborative effort by all the authors. Saud Alzahrani, and Thamer Alghamdi handled the study's planning, data entry, and statistical analysis. Additionally, they have written the results part as well. Adnan Alessa contributed to the writing of the methodology and discussion. Also, Yahya Almarhabi, Hasna Alghamdi and Abdullah Alamri carried out the final draft of the manuscript editing and revision. Omar Alshaikhi, Hatem Alessa and Badr Almaedy was in charge of the literature review and drafting of the introduction. The general supervisor of this work is Dr Mohamed A. Elhefny, who offers directions and corrections.

Funding

This study has not received any external funding.

Conflict of interest

The authors declare that there is no conflict of interests

Data and materials availability

All data associated with this study are present in the paper.

REFERENCES AND NOTES

1. Memish ZA, Saeedi MY. Six-year outcome of the national premarital screening and genetic counseling program for sickle cell disease and β-thalassemia in Saudi Arabia. Ann Saudi Med 2011; 31 (3): 229–35. doi: 10.4103/0256-4947.81527.
2. Al-Aama JY. Attitudes towards mandatory national premarital screening for hereditary hemolytic disorders. Health Policy (New York) 2010; 97 (1): 32–7. doi: 10.1016/j.healthpol.2010.02.009.
3. Alenazi SA, Ali HW, Alharbi MG, Alenizi AF, Wazir F. Prevalence of thalassemia and sickle cell disease in northern border region of Saudi Arabia. Kashmir J Med Sci 2015; 1 (1): 3–6.
4. Al Hamdan NA, AlMazrou YY, AlSwaidi FM, Choudhry AJ. Premarital screening for thalassemia and sickle cell disease in Saudi Arabia. Genetics Med 2007; 9 (6): 372–7. doi: 10.1097/GIM.0b013e318065a9e8.
5. Al-Khaldi YM, Al-Sharif AI, Sadiq AA, Ziady HH. Attitudes to premarital counseling among students of Abha Health Sciences College. Saudi Med J 2002; 23 (8): 986–90.
6. Alotaibi MM. Sickle cell disease in Saudi Arabia: A challenge or not. J Epidemiol Glob Health 2017; 7(2): 99.
7. Alsaeed ES, Farhat GN, Assiri AM, Memish Z, Ahmed EM, Saeedi MY, Al-Dossary MF, Bashawri H. Distribution of hemoglobinopathy disorders in Saudi Arabia based on data from the premarital screening and genetic counseling program. J Epidemiol Glob Health 2017; 7(S1): S41. doi: 10.1016/j.jegh.2017.12.001.
8. Amr MAM, Amin TT, Al-Omair OA. Health related quality of life among adolescents with sickle cell disease in Saudi Arabia. Pan African Med J 2011; 8 (1).
9. Amoudi AS, MBBS, Balkhoyor AH, Abulaban AA, Azab AM, Radi SA, Ayoub MD, MBBS, Albayrouti BT. Quality of life among children with beta-thalassemia major treated in Western Saudi Arabia. Saudi Med J 2013; 34 (12): 1281–6.

10. Bakri MH, Ismail EA, Elsedfy GO, Amr MA, Ibrahim A. Behavioral impact of sickle cell disease in young children with repeated hospitalization. Saudi J Anaesth 2014; 8 (4): 504.
11. Binshihon SM, Alsulami MO, Alogaibi WM, Mohammedsaleh AH, Mandourah HN, Albaity BS, Qari MH. Knowledge and attitude toward hemoglobinopathies premarital screening program among unmarried population in western Saudi Arabia. Saudi Med J 2018; 39 (12): 1226.
12. Bittles AH, Grant JC, Sullivan SG, Hussain R. Does inbreeding lead to decreased human fertility? Ann Hum Biol 2002; 29 (2): 111–30.
13. Colah R, Surve R, Nadkarni A, Gorakshakar A, Phanasgaonkar S, Satoskar P, MohantyD. Prenatal diagnosis of sickle syndromes in India: dilemmas in counselling. Prenatal Diagnosis: Published in Affiliation with the International Society for Prenatal Diagnosis 2005; 25 (5): 345–9.
14. Hasan SP, Hashmi S, Alhassen M, Lawson W, Castro O. Depression in sickle cell disease. J Natl Med Assoc 2003; 95 (7): 533.
15. Kahhaleh F, Sulaiman MA, Alquobaili F. Association of Xmn1 polymorphism and consanguineous marriage with fetal hemoglobin in Syrian patients with sickle cell disease. Cogent Med 2019; 6 (1): 1639243.
16. Kumkhaek C, Taylor JG, Zhu J, Hoppe C, Kato GJ, Rodgers GP. Fetal haemoglobin response to hydroxycarbamide treatment and sar1a promoter polymorphisms in sickle cell anaemia. Br J Haematol 2008; 141 (2): 254–9.
17. Madani BM, al Raddadi R, al Jaouni S, Omer M, al Awa MI. Quality of life among caregivers of sickle cell disease patients: a cross sectional study. Health Qual Life Outcomes 2018; 16 (1): 1–9.
18. Memish ZA, Owaidah TM, Saeedi MY. Marked regional variations in the prevalence of sickle cell disease and β-thalassemia in Saudi Arabia: Findings from the premarital screening and genetic counseling program. J Epidemiol Glob Health 2011; 1 (1): 61–8. doi: 10.1016/j.jegh.2011.06.002.
19. El-Mouzan M, Al-Salloum AA, Al-Herbish AS, Qurachi MM, Al-Omar AA. Regional variations in the prevalence of consanguinity in Saudi Arabia. Saudi Med J 2007; 28 (12): 1881–4.
20. Nwogoh B, Ofovwe C, Omoti C. Health-related quality of life in sickle cell disease subjects in Benin City, Nigeria. Afr J Med Health Sci 2016; 15 (2): 80.
21. Theodoridou S, Alemayehou M, Prappas N, Karakasidou O, Aletra V, Plata E, Tsaftaridis P, Karababa P, Boussiou M, Sinopoulou K, Hatzi A, Voskaridou E, Loutradi A, Anna Manitsa A. Carrier screening and prenatal diagnosis of hemoglobinopathies. A study of indigenous and immigrant couples in northern Greece, over the last 5 years. Hemoglobin 2008; 32 (5): 434–9.
22. Wallen GR, Minniti CP, Krumlauf M, Eckes E, Allen D, Oguhebe A, Seamon C, Darbari DS, Hildesheim M, Yang L, Schulden JD, Kato GL, VI JGT. Sleep disturbance, depression and pain in adults with sickle cell disease. BMC Psychiatry 2014; 14 (1): 1–8.
23. Wonkam A, Ngo Bitoungui VJ, Vorster AA, Ramesar R, Cooper RS, Tayo B, Lettre G, Ngogang J. Association of variants at BCL11A and HBS1L-MYB with hemoglobin F and hospitalization rates among sickle cell patients in Cameroon. PLoS One 2014; 9 (3): e92506.