Knowledge and Attitude of Families of Children with Hemoglobinopathies Toward Pre-Marital Testing in Yemen: A Pilot Study

معرفة وسلوك العوائل الأطفال المصابين بتواتر اعتلالات الهيموغلوبين لفحص ما قبل الزواج باليمن : دراسة أولية

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ملخص البحث:

يلعب فحص ما قبل الزواج دور مهم في السيطرة على الكثير من الامراض الوراثية كاعتلالات الهيموقلوبين والذي يعتبر زواج الاقارب من اهم العوامل لحدثوها. هدفت هذه الدراسة لاستكشاف معرفة وسلوك عوائل مرضى اعتلالات الهيموقلوبين خاصة الثلاسيميا وفقر الدم المنجلي لفحص ما قبل الزواج وذلك من خلال المقابلة المباشرة من قبل طبيب الاطفال لأهالي الاطفال المصابين بتواتر اعتلالات الهيموغلوبين المراجعين لعيادة امراض الدم في مستشفى الامومة والطفولة في المكلا.

شملت هذه الدراسة ٤٣ حالة من اهالي الاطفال المصابين بتواتر اعتلالات الهيموغلوبين. واظهرت ان نسبة زواج الاقارب 86.4٪ من الدرجة الاولى. و25٪ فقط يعرفوا فحص ما قبل الزواج للحد من حدوث هذه الامراض. ويعتقد 56.8٪ في تقبل المجتمع لهذا الفحص وذلك من خلال اللقاء والشرح المباشر مع الناس. وان 54.5% لا تعرف الوقت المناسب لعمل هذ الفحص. وبالرغم من انهم يشجعون اطفالهم لعمل الفحص بنسبة ٥٦,٨٪ الا ان 34.1 ٪ فقط مع انفصال الخطيبين إذا كانا حاملين للمرض. كما ان ٣ حالات فقط من مجموع الحالات تحبذ اجهاض الجنين إذا كان مصابا بأحد هذه الامراض.

تبين هذا الدراسة قلة المعرفة بفحص ما قبل الزواج لاعتلالات الهيموقلوبين الا انها تظهر سلوك ايجابي نحوه. وعليه تؤكد هذه الدراسة على انشاء برنامج الوقاية من الامراض الوراثية خصوصا اعتلالات الهيموقلوبين واتخاذ السبل الكفيلة لإنجاحه.

الكلمات المفتاحية: فحص ما قبل الزواج، اعتلالات الهيموقلوبين، معرفة وسلوك، اليمن

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ABSTRACT:

Premarital screening plays an important role in the control of genetic disorders such as Hemoglobinopathies in which consanguineous marriage increases their incidence. The aim of the study was to explore the knowledge and attitude of premarital testing among the families of affected children with thalassemia and sickle cell disease. The study was done through a direct interview with family members attending hematology clinic using a designed questionnaire.

The study involved 43 family members. It showed a high consanguineous marriage with 86.4% of first degree. Only 25 % knew about premarital testing. However, 56.8% of the participants thought that the test could be accepted by the population through direct meeting and discussion. As many as 54.5% had no idea about the best time of the test. Despite 56.8% would encouraged their offspring to participate in such program, 34.1% in favor of separation of the couples if they are carriers, and only 3 out of the 43 would go for termination of pregnancy if they had affected fetus

This study showed a lack of knowledge about the premarital testing, but a positive attitude toward it. It stresses the importance and initiation of preventive program of the most common genetic disorders particularly hemoglobinopathies.

To our knowledge no similar previous study has been carried out in Yemen .

Key words: *Premarital screening, Haemoglobinopathies, Knowledge & Attitude, Yemen.*

Introduction:

Hemoglobinopathies are considered the most common genetic disorder worldwide (1), of which thalassemias and sickle cell disease (SCD) are the most common (2) causing anemia, severe pain, stroke, susceptibility to life-threatening infections, and cardio-pulmonary compromise. Their Chronicity, severity and the high cost of life-long treatment make them a major public health problem which necessitates an active measure to control them (2).

Hemoglobinopathies are a group of inherited recessive disorders of the hemoglobin (Hb) molecule. The thalassemias in particular are quantitative Hb disorders which characterized by a defect in the synthesis of the polypeptide chains (globins) needed to assemble the Hb tetramer, while sickle cell disease is qualitative Hb disorder in which a transversion in the second residue of codon 6 of the beta-globin gene, resulting in the replacement of glutamic acid by valine (1).

The World Health Organization (WHO) highlight the importance of community control and prevention through screening program for hemoglobinopathies as an intervention that would help in detecting carrier states (3). Such screening program can be carried out by simple techniques such as electrophoresis for both prospective partners (male and female). The couple can then choose whether or not to have an affected child. Such

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program has been developed in many countries in the Mediterranean and Arabian Gulf regions which are based on carrier screening and counseling of couples at marriage (4-7). These programs are of considerable importance to decrease the incidence and prevalence of congenital and genetic disorders. High risk family oriented screening program is preferred to the whole population in communities with a high level of consanguinity and large families for identifying at risk of producing affected children (8). The success of these programs depends on adequate religious support, governmental policy, education and Counseling (9).

Yemen lacks such programs because of economical, technical, ethical issues, lack of laws to support termination of pregnancy when the disorder is diagnosed early enough in pregnancy, in addition to the social practices which support and prefer consanguineous marriage. The paediatric hematology clinic at Mukalla maternity and Children hospital is the main clinic serves the three governorates (Hadramout, Mahra, and Shabwa) of Yemen. The majority of children with Hb disorders are treated and followed up at least for some time in this hospital.

In this study we intended to explore the knowledge and attitude of premarital testing among the families of affected children with thalassemia and sickle cell disease. At the time of the study there are about 89 children with Hb disorders treated and followed up at our paediatric haematology clinic.

Material and Methods:

This is a pilot descriptive study involved 43 family members of affected children with hemoglobinopathies attending the paediatric haematology clinic at Maternity & Children hospital in Mukalla/Hadramout/Yemen. The family members were those who brought the affected child to the clinic for treatment and follow up. There is one clinic per week and the study conducted during the period from Jan 2017 to Apr 2017. Data were collected during interview by paediatrician using prepared questionnaire designed by the author. The questionnaire was revised by paediatric team where some questions modified and others included such as the best time of performing the test, and the issue of termination of pregnancy in case of affected fetus. The interview lasted about 8 minutes.by a paediatrician who explained the question to participants if needed. The questionnaire included the Hb disorders of the child, characteristics of the family member such

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number of children in the family, educational level and economic status. The knowledge and attitude toward premarital testing included acceptance and rejection, timing, attitude toward positivity of the test. The economic status was divided into 3 level according to the family monthly income in Yemeni Riyal: low if the income less than 39,000 Yemeni Riyals (YR), middle from 40,000-100,000 YR, high if more than 100,000 YR.

All data were analyzed and summarized in the form of frequencies, percentages using statistical package for social sciences (SPSS) version 22.

The participants were told they are free to participate and it would not affect their children's management.

The hospital director approved the study through the consultant paediatrician who is in charge of the clinic.

Results:

There were 26 (63.6%) thalassemias, 10 (27.7%) sickle cells and 5 cases (11.4%) sickle-thal.

Table 1 summarized the background characteristics of the participants. Nearly 43% were of low economic status, and 50% of the parents had a minimum of 5 children from which 1 or 2 were affected with either of these disorders.

There was a high consanguineous marriage of 41 (93.2%) of which 38 (86.4%) were first degree (Figure1).

Figure 2 showed the education level of the participants in which 43.2% were illiterate and neither of the participants were of higher education level.

Table 2 showed Frequency and percentage of participants to items in the questionnaire. The concept of the genetically inheritance of these disorders were recognized by 32 (72.7%). Eleven (25%) knew the premarital test as a preventive measure.

Figure 3 showed that twenty seven participants (61.4%) thought that people ignorance is the main obstacle of doing the premarital screening and 11 (25%) thought of religious reason.

Twenty five (56.8%) of the participants thought that the public would accept the premarital screening through direct meeting and discussion. The majority of participants 24 (54.5%) had no idea about the best timing of the

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test. Nearly 57% would encourage their un-affected children to participate in such program, and fifteen (34.1%) of the participants in favor of separation before marriage if premarital testing resulted of both engaged couple were carriers, while 17 (38.6%) against separation, and 11 (25%) had no idea. Three family members (6.8%) out of the 43 family members accepted termination of pregnancy if they had affected fetus.

Table 3 compared Consanguinity rates and first cousin marriages in some countries of the Eastern Mediterranean Region to our result. In this study, the consanguinity rate was 93.2% and 86.2% of first degree.

Discussion:

Genetic disorders pose a remarkable health burden on the affected families and in country with limited resources. The impact is even higher in communities with a high prevalence of consanguineous marriages and large families. Consanguinity is the important factor that contributes to the high prevalence of genetically determined disorders, because it increases the risks of recessively inherited diseases (1). The consanguinity rates in Eastern Mediterranean countries range from 16.5% to 55% (10), and the chance in a first-cousin marriage of having an affected child is considerably greater than in the case of unrelated parents. In this study the proportion of consanguinity and first cousin marriages (93.2%, 86.4% respectively) was higher compared to other Arab countries as Iraq (57.9%, 30%) Egypt (29%, 11.4%), Kuwait (54.3%, 30.2%), and Saudi Arabia (55%, 31.4%), and even higher than that found by Al-Kherbash et al among thalassemic patients in Sana'a/Yemen who found the consanguinity rate is 74.2% and 64.2% of first-degree relatives (11). The reason of such high proportion of consanguinity in our study, in one hand might be because of the small sample and hospital based of the families of affected children with these disorders, and on the other hand the majority of the participants were of lower education level (43.2% illiterate, 13.6% just read and write) and we think such group will abide the family support and preference of consanguineous marriage. We think the consanguinity proportion would be lower if a large sample size and community based study was conducted.

Several countries in the region have introduced premarital screening program for haemoglobinopathies and have made it mandatory. For example in Saudi Arabia the third Royal Decree was issued in 1/1/1425H; 21/2/2004G, for the compulsory application of pre-marital screening as

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means of preventing the most common genetic diseases (12), the same thing in Iran (13) and Turkey (14). However, experience shows that if options are not made available to carrier couples such programs will not be effective in reducing the burden of genetic diseases, as many carrier couples go ahead with their planned marriage (15).

The acceptance of such preventive programs in Muslim communities had been hampered by many factors such as religious beliefs, cultural norms, traditions, literacy and education level social, and education (16). In our study, the participants thought the main obstacle against acceptance is people ignorance (61.4%) rather than religious reason (25%) and people would accept the test through direct meeting and discussion (Fig 3).

In Saudi Arabia experience, despite of the positive attitude toward the premarital screening test, fears were expressed towards the confidentiality of results and it was felt that social and psychological problems would ensue from abnormal results. (17). such concern has to be put in mind before applying such program.

The aim of premarital screening program is to decrease the incidence of hemoglobinopathies by counselling the carrier couples and the decision to marry is then left to them. In our study the attitude towards cancellation of marriage was 34.1%. Reports from other countries showed the incidence of cancellation of marriage if both couple were carriers for hemoglobinopathies had been raised, for example in Saudi Arabia more than 5-fold increase between 2004 and 2009 (from 9.2% to 51.9% (18), in Greece and Italy about 80% [19], and the highest was in Iran as reported by Ghanei et al 90% (20). Discouraging marriages particularly between cousins is neither feasible nor desirable particularly in our region (12) and this has to be considered. So we think the result of the tests to couples, if positive, should have no impact on the decision to proceed with the marriage or not. However, they should offered counseling appropriately by trained personnel about the complexity of these disorders, and implication on their future children.

Acceptability of termination of pregnancy is based on religious and social grounds (21). In Saudi Arabia despite a 1990 ruling (Fatwa) allows termination of pregnancy in the first 120 days after conception if the fetus is shown beyond doubt to be affected with a severe malformation that is not amenable to treatment (22), Muslims may reject prenatal diagnosis on religious grounds for the pregnancy termination,

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The majority of our participants (54.4 %,) had no idea about the best timing of performing the premarital screening of hemoglobinopathies (table 2). However, in Italy, it is preferred before leaving school (23). Others suggested in high school to identify the carrier states before the timing of marriage and doing the premarital screening as the last stage of the marriage arrangements poses a major stress and embarrassment for both families to break all arrangements of the marriage (24). Others proposed that screening singles on admission to university prior to any commitment may be preferable than screening immediately before the marriage certificate is issued (25). In Yemen because many students leave school early, it might be worth to do early during school period so as to catch the carriers before marriage arrangement.

Conclusion:

Our study provided, for the first time an overview of importance of the premarital screening program for hemoglobinopathies in Yemen. Despite lacking of knowledge, the participants showed a positive attitude toward premarital screening program. The program is cost effective preventive measure, and for successful implementation certain elements should be taken into account such as appropriate health education of the public, trained personnel in counseling high risk couples, confidentiality of addressing the positive result and timing of screening in relation to marriage.

Recommendation:

- The department of health should initiate preventive measures through the premarital program and screening for thalassemia and sickle-cell disease.
- Involve all community leaders particular the religious one to clarify and correct misunderstanding of Islamic rules.
- liaise with the health institutions and organizations such as Thalassemia and Sickle Cell Anemia Society at the national level and internationally.
- Further community based study should be conducted.

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Limitation of the study:

- Hospital based of selected group (family of affected children with haemoglobinopathies).

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Table 1: Characteristics of study population:

Characteristic	No	Percentage %
Hemoglobin disorder:		
Thalassemia	28	65.1
Sickle cell disease	10	23.3
Sickle thal	5	11.6
Family member (interviewee):		
Fathers	20	45.5
Mothers	18	40.9
Grandfathers	2	4.5
Grandmothers	3	6.8
Others	0	
Education level of family member:		
Illiterate	19	43.2
Read and write	6	13.6
Essential Education	9	20.5
Secondary	9	20.5
University	0	0

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Economic status:		
Low	19	43.2
Middle	18	40.9
High	6	13.6
Total no. of children in the family		
Less than 5	21	47.7
5 – 10	21	47.7
More than 10	1	2.3
Affected children in the family:		
1	22	50
2	20	45.5
3 and more	1	2.3
Consanguinity:		
First degree	38	86.4
Second degree	3	6.8
Non consanguineous	2	4.5

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Table 2: Frequency and percentage of participants to items in the questionnaire (N = 43):

Questionnaire	No	Percentage (%)
How can either of the diseases are prevented:		
o Premarital testing	11	25
o Don't get married to a relative	9	20.5
o Nothing as it is God wish	9	20.5
o I don't know	14	31.8
What are the main Obstacles of doing premarital testing:		
o People ignorance	27	61.4
o Religious reason	11	25
o Lack/limit of health education	4	9.1
o Others		
o Don't know	1	2.3
How to make people accept pre-marital testing		
o Through different media	14	31.8
o Through religious scholars	4	9.1
o Meeting directly and discussion with people	25	56.8
When do you think the best time to perform the test		
o During secondary school	10	22.7
o At the time of engagement and before complete marriage	9	20.5
o No idea	24	54.4
Would you encourage your children to go for pre-marital testing:		
o Yes	25	56.8
o Don't know	18	40

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If both partners are carriers of the disease, would you prefer their separation before marriage:		
o Yes	15	24.1
o No	17	38.6
o Don't know	11	25

Table 3: Consanguinity rates and first cousin marriages in some countries of the Eastern Mediterranean Region including our study:

Country	Total consanguinity Rate (%)	% first cousin marriages
Saudi Arabia	55	31.4
Egypt	29	11.4
Kuwait	54.3	30.2
Iraq	57.9	30
Yemen (This study)	**93.2**	**86.4**

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Figure 1: Frequency of the parent consanguinity among the studied cases (No 43)

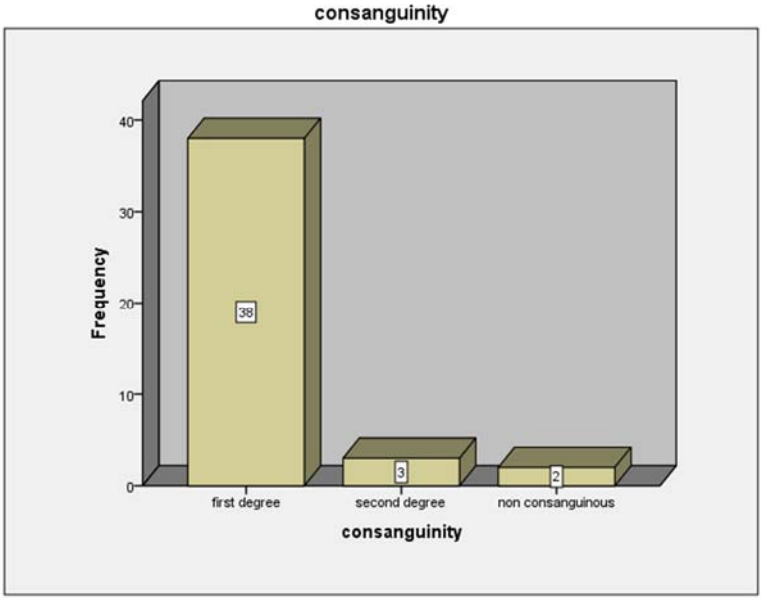

Figure 2: frequency of education level among the studied cases

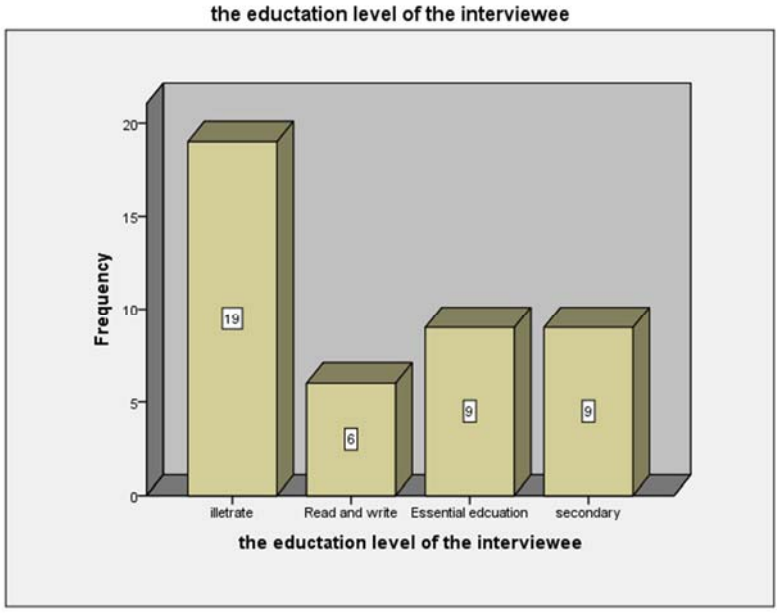

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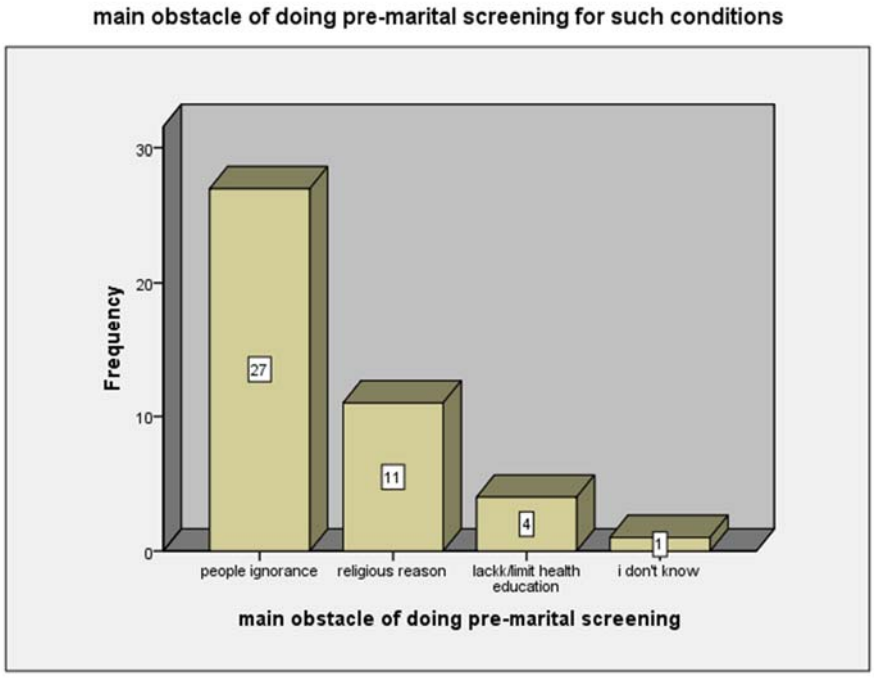
Figure 3: Frequency of the main obstacles of doing pre-marital screening among the studied cases (No 43)



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