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RESEARCH ARTICLE

Public education for the prevention of hemoglobinopathies: a study targeting Kocaeli University students

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ABSTRACT

In addition to premarital screening programs, education of the general population is important in preventing hemoglobinopathies. The aim of the present study was the education of university students. Short questionnaires were applied before and after a prepared lecture. A 20-minute audiovisual education was provided including the clinical characteristics and inheritance of thalassemia and sickle cell anemia (SCA) as well as the importance of carrier screening. The attendance to 42 lectures was low (n= 1348 = 5%). Seventy-four percent of the students had heard of thalassemia. The percentage of the students who had some accurate knowledge was 25% for thalassemia and 11.7% for SCA. Following the lecture, these numbers increased to 86.2% and 72.1% for thalassemia and SCA, respectively. Only 13.6% of the students were aware of the risk of being carriers, but this increased to 78.6% following the lecture; 92.4% learned that these hemoglobinopathies were inherited and 78.8% learned that consanguineous marriages would increase the risk. Education on hemoglobinopathies must be integrated into the curricula of middle schools and high schools.

Key Words: Thalassemia, sickle cell disease, prevention, education

ÖZET

Hemoglobinopatilerin önlenmesinde toplumun eğitimi: Kocaeli Üniversitesi öğrencilerini hedef alan bir çalışma

Hemoglobinopatilerin önlenmesinde evlilik öncesi tarama programları kadar toplumun eğitilmesi de önemlidir. Bu çalışmada üniversite öğrencilerinin hemoglobinopatiler konusunda bilgilendirilmesi amaçlanmıştır. Eğitim öncesinde ve sonrasında öğrencilere açık ve kapalı uçlu sorular içeren anket verilmiştir. Barkovizyon kullanılarak talasemi ve orak hücreli aneminin klinik özelliklerini, genetik geçişini, kan sayımı ve hemoglobin elektroforezi ile yapılan taramanın hastalığın önlenmesindeki önemini anlatan 20 dakikalık eğitim verilmiştir. Yapılan 42

seminerde katılım oranı düşüktür (n: 1348, %5). Katılımcıların %74'ü talasemiyi daha önce duyduğunu belirtmekle birlikte bunların sadece %25'i, %11.7'si orak hücreli anemide doğru bilgi sahibiydi. Eğitim sonrası talasemide ve orak hücreli anemide bu oranlar %86.2 ve %72.1'e yükseldi. Katılanların sadece %13.6'sı taşıyıcı olmanın riskini bilirken bu oran eğitimle %78.6'ya yükseldi. Eğitim sonrası öğrencilerin %92.4'ü bu hastalıkların kalıtımla geçtiğini, %78.8'i akraba evliliklerinin hasta çocuğa sahip olma riskini arttırdığını öğrendi. Hemoglobinopatilerin önlenmesi isteniyorsa bu konular orta ve yüksek öğretimin ders programlarında yer almalıdır.

Anahtar Sözcükler: Talasemi, orak hücreli anemi, önleme, eğitim

INTRODUCTION

Thalassemia is a recessively inherited hemoglobinopathy characterized by chronic anemia. It requires monthly blood transfusions and regular iron chelation. It is a real burden for the individual, the patient's family and the health care system[1]. In 2003, approximately 25% of the national blood products and 6 million vials of deferoxamine were used for thalassemic patients in Iran, and in the Maldives, 60% of the entire blood requirement of the country was used for the treatment of thalassemic patients[2,3].

Hemoglobinopathies are also important health problems in Turkey. Many local screening studies were performed before a national policy of prevention was established. Regulations for the Prevention of Hereditary Blood Disease (hemoglobinopathy control program) were published in 1993. In 1994, the Ministry of Health established Thalassemia Centers and started premarital screening in the south of the country (Antalya, Antakya, Mersin, and Muğla), where the prevalence of severe hemoglobinopathies was high. In the succeeding years, the prevention program was broadened and premarital screening centers became active in 33 cities. The results of the screening studies covering 16 cities and 377.339 healthy people showed a prevalence of 4.3% for beta-thalassemia trait. Antalya had the highest prevalence of beta-thalassemia trait (13.1%) and Cukurova region had the highest rate for HbS trait (10%)[4].

Two hundred and fifty million people, 4.5% of the world population, carry a pathological hemoglobin gene. Each year 300.000 infants are born with major hemoglobinopathies. Recommendations of the World Health Organization are followed in many countries[5]. In 1974, public education, population screening for heterozygotes, genetic counseling for carrier couples, mandatory premarital screening and antenatal diagnosis were started as components of a prevention program in Cyprus. There has been a sharp fall in the number of babies born with beta-thalassemia disease. In 1974, 53 of 6594 births were homozygote, and after 1986, only five homozygote babies were born[6].

To estimate the prevalence of sickle cell disorders and beta-thalassemia, with their regional distribution, an adult population was screened as part of the Saudi Premarital Screening Program between February 2004 and January 2005. Among the 2.375 high-risk couples contacted by telephone, 89.6% married each other despite the known high-risk status. They concluded that "The results showed excellent access to the target population". However, the program's objective of decreasing high-risk marriages was not as successful, indicating the need for improvement in the health education program for the public, more efforts in counseling high-risk couples, and changes in the strategy of timing of screening in relation to marriage[7].

Many studies show that in addition to screening of heterozygotes, education of the population is essential for prevention of homozygote births.

MATERIALS and METHODS

In the present study, performed between October 2004 and May 2005, the aim was the education of Kocaeli University students about hemoglobinopathies and evaluation of their

knowledge on the subject before and after an educational lecture. Consents of the University rector and deans of the faculties were obtained. The study was announced in every school with posters at least one week before the lectures. For the study, a short questionnaire with multiple choice and open-ended questions were applied to evaluate the students' awareness about the subject. Audiovisual education about the clinical characteristics and inheritance of thalassemia and sickle cell anemia (SCA) and the importance of carrier screening with blood counts and hemoglobin electrophoresis lasted 20 minutes and was presented by a last-year resident in pediatrics. The efficacy of the education was evaluated by a short questionnaire form after the lecture. A printed material was also distributed to the participants.

Pre-Lecture Questionnaire

1. Have you ever heard about a disease called thalassemia (Akdeniz anemisi)?
   a. I know it from TV
   b. I heard on the radio
   c. I read in the newspaper
   d. I heard at school
   e. I have a relative with this disease
   f. I know someone with this disease
   g. I have a relative who is a thalassemia carrier
   h. I have never heard
   i. I learned from another source

2. If you have heard about thalassemia, write what you know in a few sentences.

3. Have you ever heard about a disease called sickle cell anemia?
   a. I know it from TV
   b. I heard on the radio
   c. I read in the newspaper
   d. I heard at school
   e. I have a relative with this disease
   f. I know someone with this disease
   g. I have a relative who is a thalassemia carrier
   h. I have never heard
   i. I learned from another source

4. If you have heard about sickle cell anemia, write what you know in a few sentences.

5. Are these diseases inherited?
   a. Yes, inherited c. I have no idea
   b. No

6. Do you know the importance of being a carrier of these diseases?
   a. I don't know
   b. If you know, explain

7. Do you know if you are a carrier for any of these diseases?
   a. I don't know
   b. If you know, write if you are a carrier of thalassemia or sickle cell anemia

8. Is it possible to identify carriers?

Post-Lecture Feedback Questionnaire

1. What are the main characteristics of thalassemia (Akdeniz anemisi)? What are the complaints of the patients? What is the treatment?

2. What are the characteristics of sickle cell anemia? What are the complaints of the patients?

3. Are these diseases inherited?
   a. Yes, inherited c. I have no idea
   b. No

4. What is the impact of consanguineous marriages on these diseases?

5. What is the impact of being a carrier of these diseases?

6. Do you want to have a blood test to learn if you are a hemoglobinopathy carrier?
   a. Yes b. No

7. What is your opinion about the study?

RESULTS

There were a total of 42 lectures scheduled and although dates of the lectures were chosen after interviews with administrative offices of the schools, the attendance was very low (1348/26840= 5%).

Evaluation of The Questionnaires

Pre-lecture questionnaire: Although 74.4% stated that they had heard of thalassemia, only 18.2% (n= 245) knew that it is an anemia, 3.3% (n= 47) knew that it is an inherited disease, and 3.5% (n= 47) knew that it is an inherited anemia. Of all the participants, 12.7% heard of thalassemia on TV, 14.2% at school, 13.5% on TV and in a newspaper, 16.4% on both TV and at school, 5.8% did not answer the question, and the rest had heard from other sources.

Of all the participants, only 1.6% had heard of SCA on TV, 15% at school, 0.7% on TV and in a newspaper, 3.2% on both TV and at school, and 9.3% did not answer the question. Although 90.7% said that they had heard of SCA, only 9.7% (n= 131) knew that it is an anemia, 1.3% (n= 18) knew that it is an inherited disease, and 0.7% (n= 10) knew that it is an inherited anemia.

Some students thought that thalassemia was a cancer of blood cells, a type of epilepsy, a contagious disease, a bleeding disorder, or a result of iron or vitamin B12 deficiency.

Of all the participants, 41.5% answered that these diseases are inherited, but only 13.6% of them were aware that carriers would transfer the defective gene to the new generation. Three students answered that they had the thalassemia trait and one student had thalassemia major.

Post-lecture feedback: Of all the participants, 78.6% (n= 1062) learned the importance of being a carrier, 92.4% (n= 1045) learned that these hemoglobinopathies were inherited and 78.8% (n= 1062) learned that consanguineous marriages would increase the risk of having a sick child. Eighty-six percent of the students expressed wanting to be screened by blood tests, 90.2% explained that the lecture was very useful, 6.8% thought that similar lectures about health should be performed in schools, 0.8% (n= 11) wanted to inform others on the subject, and 5.6% suggested presentation of these lectures to the public as well.

DISCUSSION

This study highlights the lack of knowledge about hemoglobinopathies among university students, although there is a high prevalence of thalassemia and SCA in Turkey. A similar study was performed in high schools of Antalya in 1998-1999, where prevalence of thalassemia is very high. Only 10.7% of the seniors were informed of the inheritance of thalassemia and the results of a marriage of two carriers[8]. An educational study was performed in 8th grade classes of Cukurova, where prevalence of hemoglobinopathies is also high, and its efficiency was reported[9].

Open-ended questions instead of multiple choice were preferred for this study population of high school graduates, who could express their knowledge in writing, in an effort to prevent correct responses by chance.

A control program for an inherited disorder is defined as an integrated strategy combining the best possible patient care with prevention by community education, population screening, genetic counseling and the availability of prenatal diagnosis[10,11].

A study from south of Turkey evaluated the outcomes of premarital screening studies. Marriage of two carriers was reported in 1.2% (n= 120) of screened couples. Ninety-three of these couples were enrolled. Five decided not to marry and one couple divorced. Despite premarital genetic counseling, 35.6% (n= 31) were unaware of being a carrier. Of the 56 couples who knew that they were carriers, 43 knew the impact of being a carrier and 38 knew about prenatal diagnosis. During the survey, 63 couples were pregnant but only 11 (17.5%) had undergone prenatal diagnosis. The study showed that without public education, premarital screening studies cannot fully prevent birth of homozygote babies[12]. People perceive premarital hemoglobinopathy screening as completing one of the official documents and rarely decide not to marry, and they frequently cannot understand counseling because they do not have any basic knowledge.

Performing educational sessions and screening of 11th grade students, as in Bahrain, would be more logical to give them a better chance of choosing a suitable partner. One of the students in the present study wrote that it was late for him to be informed about hemoglobinopathies. Although it is not as disastrous as learning about being a carrier during antenatal visits, it is still distressing for the engaged couples and their families to learn that they are carriers just before marriage. As a part of the National Student Screening Project to determine the prevalence of genetic blood disorders and to raise awareness among young Bahrainis, they screened 11th-grade students, organized lectures and distributed information about these disorders[13]. A similar program is also successfully organized for all students 14-18 years of age in Montreal, Canada[14].

Although termination of early pregnancy is legal in our country, religious and ethical considerations also prevent prenatal diagnosis and lead to birth of homozygous children. Due to religious restrictions on abortions in Isfahan-Iran, premarital screening could not prevent the birth of homozygote babies. They started education covering marriage officers, health authorities and imams. Pre-marriage classes, broadcasts on TV and published leaflets were used to spread information. As a result, 90% of the high-risk couples decided not to marry. This approach seems rational for some Islamic countries and countries with limited health budgets. They recommend annual screening of students of a specific grade and of school dropouts in order to give carriers the choice of a non-thalassemia carrier partner[15].

Some other countries also have a problem with hemoglobinopathy control. In China, the frequency of carriers for alpha-thalassemia is 15% and beta-thalassemia carriers comprise 4.8% of the population. Among 106 patients with beta-thalassemia major followed in a clinic, the majority died before five years of age. Knowledge surveys about thalassemia were conducted. Their results showed a severe lack of knowledge about thalassemia among both medical professionals and the general population. They concluded that there was an urgent need to educate the general population and the medical community for a successful community-based prevention program[16].

The importance of public information was well illustrated in a Sardinian program, where a sharp rise in affected births was observed after funding for public education was discontinued. In Sardinia, carrier screening is carried out voluntarily for couples of child-bearing age with or without pregnancy. The education of the population is achieved via mass media (TV and newspapers) and meetings at high schools, industries (factories) and large shops. Special educational meetings are periodically organized to inform general practitioners, obstetricians, paramedical professionals and midwives. They comment that the high efficiency of their program depends on the extended-family examination of any person detected with heterozygous or homozygous beta-thalassemia[17].

In addition to its economic and physical burden, thalassemia is a real psychosocial burden for the patients and their families. Ninety-three percent of the families say that they would have chosen to terminate the pregnancy if they had known the fetus had thalassemia major[1]. However, before having a child with thalassemia and experiencing the problems, a family's motivation is not as high to terminate the pregnancy due to their lack of knowledge. Previous studies in Europe[18] showed that the majority of homozygous births were due to lack of information (81%), but few were due to informed parental choice (10%) and diagnostic errors (9%).

Although premarital screening is an obligation in 33 cities of our country, there is no national strategy for education of the population about the subject. It is obvious that premarital consultation is difficult because most people know little of genetics. In 2007, the Thalassemia Federation organized a mobile education center to travel throughout the country targeting public and local physicians.

In Greece and Crete, interviews of 35.000 people aged 15-65 years were performed to investigate public awareness of the basic characteristics of thalassemia major. Even at the lowest educational level, 89% knew that it requires blood transfusions. This reflects efforts of the Hellenic Red Cross and thalassemia association in advertising the subject and the Ministry of Health's general publicity regarding blood transfusion in the mass media. The fact that about 80% of the population who had heard of thalassemia recognized its hereditary nature suggested the effectiveness of the advertising campaigns conducted by the Ministry of Health aimed at the prevention of thalassemia[19].

Frequency of consanguineous marriages is high in our country (21.1%), ranging from 10.2% in western Anatolia to 32.9% in eastern Anatolia. First-cousin marriages are significant determinants underlying the high infant mortality[20]. In a study from the Thalassemia Center of Antalya State Hospital, the rate of consanguinity in patients with thalassemia was significantly higher than that in the population in this region of the country[21]. Risk of marriages between cousins must be emphasized during the educational programs.

It seems that expanding the premarital screening to all cities of the country is currently not possible due to the large country, high and relatively young population and young age of marriage, high rate of unofficial marriages in east Anatolia, low education level and limited budgets for preventive medicine. Nevertheless, extended-family studies throughout the country for all detected heterozygous or homozygous cases for hemoglobinopathies would be logical. This approach will be successful if post-graduate education for primary care physicians, pediatricians and gynecologists is introduced. In Pakistan, a similar approach prevented further pregnancies of high-risk couples with healthy children[22].

Maldives, with 18.1% thalassemia prevalence, initiated the "Awareness Program in Thalassemia". Extensive public education, especially in schools, contributed to thalassemia becoming a household word in the Maldives[3].

CONCLUSION

In conclusion, although the attendance was low, this study showed the effectiveness of even a short educational program on the awareness of these diseases. The importance of education in preventing thalassemia in a population where the prevalence is high is obvious. Although they might be expensive, education and screening programs can be implemented for the same cost as the annual treatment expenses of one sick child with thalassemia.

Post-graduate education of gynecologists, pediatricians, primary care physicians, midwives, teachers, school directors, imams and marriage officers. They must be obligated to attend local courses and receive certification. Visits to the hospital to observe patients with thalassemia must be included in the course program.

Collaboration of Medical Schools with regional primary care centers and state hospitals for the education of the health care professionals.

Pre-graduate education of teachers, nurses and physicians about the hemoglobinopathy control program.

Repeated educational programs in the mass media, large shops and hospital outpatient halls.

Education of young men during their military service.

Introduction of the topic of thalassemia into the curricula of middle schools, high schools and colleges.

Survey of carrier couples for repeated genetic counseling and provision of motivation for prenatal diagnosis.

Uniform videotape presentations should be used during genetic counseling in screening centers and during other education studies.

The history of patients and their parents' experiences and feelings must be recorded and used during audiovisual presentations for greater effectiveness with the public. Risks of consanguineous marriages must also be emphasized.

Informative booklets should be prepared for distribution during educational programs and genetic counseling.

Advertising campaigns should be prepared to provide public awareness. Posters with attractive photographs and remarks may be displayed in crowded avenues of the cities and some announcements may be repeated on radio and television broadcasts.

Collaboration of the Ministry of Health, Thalassemia Foundation, Medical Schools, and Ministry of National Education to establish a continuous education strategy and education team in every city.

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