An Education Programme on Sickle Cell Anemia and ß-Thalassemia for the 8th Grade Students

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ABSTRACT

ß-thalassemia and sickle cell anemia (SCA) are major health problems in Çukurova. Screening studies have been carried on in this region for many years. The government started premarital screening programme and prenatal diagnosis has been done in Çukurova University Medical Faculty since 1992. In spite of all these, the prevention programme has not been successful. The purpose of this study was to start an education programme to increase the awareness of the people for these severe hereditary diseases. The target population was the 8th grade students. A visual presentation was given to students on Thalassemia and SCA after examining the curriculum in the Biology and/or the Science books. A total of 1221 students in seven Elementary Schools were informed. A questionnaire was given to five schools before the presentation and to two after the presentation. The comparison of the two group's result showed that the information given was well received. Thus, an extensive education programme encompassing doctors, health personel, civil and religious leaders and the support of the government will result in nil SCA and Thalassemia births.

Key Words: Education, Thalassemia, Sickle cell anemia (SCA), Elemantary school.

ÖZET

Orak Hücreli Anemi ve ß-Talassemi Üzerine 8. Sınıf Öğrencileri İçin Bir Eğitim Programı

ß-talassemi ve orak hücreli anemi (OHA) Çukurova'da önemli sağlık problemleridir. Yıllardır bu bölgede tarama programları sürdürülmektedir. 1992 yılından beri Çukurova Üniversitesi'nde evlilik öncesi tarama ve prenatal tanı programı yürütülmektedir. Tüm bunlara rağmen, tarama programları başarısız olmuştur. Bu çalışmanın amacı toplumun bu ciddi herediter hastalıklara duyarlılığını arttırmaktır. Hedef kitle 8. sınıf öğrencileridir. Biyoloji ve/veya fen kitaplarındaki ders programları incelendikten sonra öğrencilere talassemi ve OHA konusunda görsel sunum yapılmıştır. Yedi ilköğretim okulunda toplam 1221 öğrenci bilgilendirilmiştir. Beş okula sunum öncesi, iki okula sunum sonrası anket yapılmış ve iki grubun sonuçlarının karşılaştırılması, verilen bilginin iyi alındığını göstermiştir. Doktorları, sağlık personelini, sivil ve dini liderleri kapsayan genişletilmiş bir eğitim programı ve devlet desteği talassemi ve OHA'lı doğumları sıfıra indirebilir.

Anahtar Kelimeler: Eğitim, Talassemi, Orak hücreli anemi (OHA), İlköğretim okulu.

Turk J Haematol 2003;20(1): 19-24

Received: *29.07.2002* ***Accepted:*** *04.11.2002*

INTRODUCTION

Thalassemia and sickle cell anemia (SCA) are the most widespread single gene disorders in the world. Each year 300.000 homozygous babies are born and it is estimated that there are 366 million carriers[1]. Prevalence studies were done in Çukurova, the Southern part of Turkey encompassing the province of Hatay, Adana and İçel. A variable frequency ranging from 0.5% to a high 30-40% for sickle cell trait and 0.1 to 10.8% for ß-thalasaemia trait in various regions with an overall carrier rate of 8% for SCA and 3% for ß-thalassemia was noted[2-5]. Turkey ranks 16th among the top 25 countries with the highest population increase. Adana and İçel have population growth above the average of Turkey due to high fertility rate and the migration from the East Anatolia. Considering the high birth rate, high illiteracy of women, consanguineous marriages above 20% and a high carrier rate, 300-400 homozygous cases are expected to be born each year[6,7]. Preventive measures by WHO are:

1. Carrier screening,
2. Education and enlightenment of the public,
3. Mutation analysis,
4. Prenatal diagnosis[8].

The regional mutations were determined by 1992 in Çukurova University (Ç.U.) Medical Faculty, Department of Biochemistry and prenatal diagnosis was started in the same year by the Prenatal Diagnostic Centre of Ç.U. Medical Faculty. The ministry of health started preventive studies with the establishment of Thalassemia Centres along with mandatory premarital screening tests in high risk areas. Though this programme began in 1994, 1998 and 1999 in Hatay, Adana and İçel, respectively, a small percentage of at risk couples go for prenatal diagnosis. Genetic counselling was given to at risk couples, however, only 11/63% requested prenatal diagnosis[5,6,9,10]. As described by Scriver, Cao, Anagnostou and Loader, for a successful control programme, carrier screening must go hand in hand with the education and enlightenment of the people[11-14]. Thus, the education and the enlightenment of the public is the missing link of the preventive programme. In this study, we present the start of an education programme designed for students in the 8th grade of the Elemantary School. In the future, this will be extended to Lycee students and in co-operation with the Office of the Ministry of Health to increase the awareness of the public, specially to emphasize the prospects of carrier status, severity, acuity of SCA and ß-thalassemia.

MATERIALS and METHODS

Selection of the Target Population

This study was done in Adana between January and June 2001. The 8th grade students in the Elementary School were chosen as the target population. The students from the periphery are mostly children of migrants from the Eastern Anatolia and of low economic status and do not continue the Lycee education. The boys start to earn a living and the girls are given in marriage. The study was conducted in 7 schools and conferences were given to a total of 1221 students.

Participants

The programme was carried out by the Adana branch of the Thalassemia association with the collaboration of Ç. U. Medical Faculty and the Office of the Ministry of Education and was supported by the Office of the Ministry of Health.

Preparation of the Education Material

The contents of the Biology and/or Science curriculum of the 8th grade students were examined. The curriculum covered basic knowledge on the cell, the nucleus, the chromosome, the composition of blood and Mendel's law of inheritance. This basic knowledge would help in the understanding of the information on the hereditary blood diseases as SCA and ß-thalassemia. A visual education programme with overhead

projection was preferred for almost all the schools had an overhead projection. Colored transparencies were prepared covering the material they had in their curriculum as given above and we described the hereditary nature of SCA and ß-thalasaemia, gave the general symptoms, showed pictures of homozygous cases emphasising the severity of the disease and the need for the life long frequent hospitilisation and blood tranfusions for ß-thalassemia, the inheritance pattern, the risks of the carrier status in marriage, and the prevention measures that are available when the couples are carriers. The programme took 35-45 minutes for the presentation and 10-15 minutes with the active participation of the students.

Questionnaire

A questionnaire was prepared and given randomly to the 8th grade students in different classes to see if the questions were asked correctly and revisions were made. The first 13 questions were for demographic data, 13-16 were multiple choice questions on the curriculum and the rest were on SCA and thalassemia. The questionnaire was given to 5 schools (Group 1), before the presentation and to two schools (Group 2) after the presentation. The results were analysed in SPSS program and $p< 0.05$ was considered statistically significant.

RESULTS

In Table 1, the demographic data of the students is given. Each percentage given in the tables was calculated with the actual numbers of respondents. As seen from the Table 1, the % of boys was greater than that of the girls especially in Bahçelievler (70%) where the students were of eastern Anatolia origin. The students from İsmet İnönü and Gazi were of upper income class and 20-30% of the parents were university graduates. The consanguineous marriages were low (10%) and the mean number of children was 2.5. Consanguinity was 44.6% and 31.8% in Bahçelievler and Kanuni schools, respectively with a mean of 19.3% (236/1221) for all the schools. The mean number of siblings was again high in these two schools, 6.3 and 5.2, respectively, with a mean of 4.1 for all. High illiteracy of women ranging 22.7-65.9% and of men from 4.5-22.1% was found in many schools though a 5 year elementary school education was compulsory for years. Many of the parents did not have social security; absence of social security ranged from a low 9.3% to a high 40% with a average of 20.1% (226/1126) for the area investigated.

The results of the questionnaire to evaluate the material covered in the curriculum and the presentation is shown in Table 2. The percentages of correct answers

Table 1. The demographic data of the students

Name of the school	N	Male %	n	c	Social security absent %	Father illiterate %	Mother illiterate %
İsmet İnönü	411	57.2	2.5	10.7	12.5	0.5	0
Gazi	252	56.0	2.5	9.9	9.3	0.1	0
Hadırlı	90	50.0	3.8	23.3	27.0	1.1	23.3
Bahçelievler	77	70.1	6.3	44.6	40.0	22.1	61.0
Cumhuriyet	218	56.4	4.3	28.4	25.0	9.6	33.5
Kanuni	88	58.0	5.2	31.8	31.0	17.0	65.9
Mithatpaşa	85	50.6	3.8	24.7	31.3	4.5	22.7
Total	1221	56.7	4.1	19.3	20.1		

N: Number of students, n: Average of siblings, c: Percentage of consanguinity.

on question (Q) 13 on the function of the nucleus were around 75% in three schools and 40% in the others.

The answers to question on hemoglobin, the choromosome and genes and anaemia were around 85% correct though anaemia was not in the curriculum. The contents of Q14-16 were in the curriculum and there was no significant difference between the percentages of correct answers before the presentation as compared to after the presentation, $p > 0.05$. Q18-22 were on ß-thalassemia and SCA and there was a significant difference between correct answers before the presentation as compared to after the presentation, $p < 0.01$. However, there was no statistically significant difference in Q23, $p > 0.05$ (Table 3).

DISCUSSION

In this study we have shown that the mean rate of consanguinity in Adana is 19.3%, similar to the mean for Turkey. However, the rate changes in different sites from 10% to a high 45%. As is well known, consanguinity increases the rate of hereditary diseases. As seen from Table 1, the high rate of consanguinity is seen in schools Bahçelievler and Kanuni where the rate of illiteracy of the parents is high. Also, the mean number of siblings in these two schools are high, 6.3 and 5.2 respectively as compared to the mean of 4.1 for all the schools. An inverse correlation is seen here between the level of education and the rate of consanguinity and the mean number of children as noted before (6.7). The choice of the 8th grade students as the target population seems to be correct in that they have the basic knowledge on haemoglobin, hereditary, the chromosome, genes and aneemia (Q14-17). As seen from Table 3, there was no significant difference in the percentage of correct answers before the presentation as compared to after the presentation. The students were receptive to the information given on ß-thalassemia and SCA in the presentation. There was a significant difference on the percentage of correct answers on SCA and ß-thalassemia after the presentation as compared to before the presentation $p < 0.01$. The results of Q23 was surprising

Table 2. Level of knowledge before the presentation and after the presentation (the rate of correct answers)

	Name of the school	Q13	Q14	Q15	Q16	Q17	Q18	Q19	Q20	Q21	Q22	Q23
G 1	İsmet İnönü	73.1	81.8	81.6	95.8	95.3	52.1	40.5	80.5	54.0	65.1	78.2
	Gazi	77.8	86.8	82.1	98.0	92.2	54.1	43.1	83.1	82.3	77.0	81.3
	Hadırlı	75.2	80.0	83.3	98.9	98.8	7.8	4.7	97.8	94.4	61.8	49.4
	Bahçelievler	46.4	56.8	55.1	87.5	59.5	27.8	32.9	50.0	49.3	52.9	72.5
	Cumhuriyet	31.4	65.5	62.8	87.7	83.5	38.5	23.4	78.5	75.6	75.4	66.7
G 2	Kanuni	37.2	94.3	68.6	96.6	95.5	69.8	71.8	88.4	45.5	97.7	83.0
	Mithatpaşa	43.6	84.7	87.1	98.8	80.0	79.5	76.5	91.8	90.5	98.8	72.6
	Total	67.0	80.1	76.7	94.9	89.5	49.4	40.3	81.6	68.4	73.4	74.3

* In the first five schools (G 1), the questionnaire response are before the presentation and in the remaining two schools (G 2) the results are after presentation.

Multiple choice questions were on:

Q13 - Function of the nucleous
Q14 - On haemoglobin
Q15 - On the chromosome
Q16 - That DNA is the site of genes
Q17 - What is anemia
Q18 - What is ß-thalassemia
Q19 - What is SCA
Q20 - Who is a carrier
Q21 - Who is homozygous
Q22 - Is thalassemia inherited
Q23 - Characteristics of SCA

Table 3. Comparison of the degree of knowledge before and after the presentation (P) in different schools

Questions	Before P corretct answer % (five schools)	After P correct answer % (two schools)	p
Q13	74.9	40.2	p< 0.01
Q14	81.3	89.6	p> 0.05
Q15	79.6	77.8	p> 0.05
Q16	96.4	97.7	p> 0.05
Average of Q13-Q16	78.7	76.3	p> 0.05
Q17	88.7	87.9	p> 0.05
Q18	40.3	74.6	p< 0.01
Q19	36.2	74.1	p< 0.01
Q20	81.6	90.1	p< 0.01
Q21	79.2	67.4	p< 0.01
Q22	67.3	89.3	p< 0.01
Q23	75.3	77.9	p> 0.05
Average of Q17-Q23	66.8	81.5	p< 0.05

P: Presentation, Q: Questions.

for the students had knowledge on SCA, that it was hereditary and that the red cells are sickle shape. This may be due to the fact that SCA is widespread in this region and the students have probably seen or heard of the sick patients[2-5].

The performance of children in schools is known to be correlated to the education and economic status of their parents. However, as seen in this study, the receptivity of children to knowledge is independent of the families' education. In both Bahçelievler and Kanuni the illiteracy of the mothers were around 60%. However, the percentage of correct answers was 53.7% in Bahçelievler before the presentation and in Kanuni 77.1% after the presentation. Our preliminary education programme seems to show that enlightenment of the students about the severity and the prevention programme of SCA and ß- thalassemia by visual presentation increased the knowledge and awareness of the children. Very many students asked if they could have their blood tested and the cost of the analysis. They would voluntarily had their blood tested if the service had been available on site.

Screening studies have been carried on in Çukurova for more than 30 years[2-5]. However, the control programme that started in 1990's have not been successful. The achievement of success in Çukurova has many hurdles. First, illiteracy of women and men are 65.9% and 23.3%, respectively in some regions and consanguineous marriages are much above the mean for Turkey as shown in Table 1. Thus, for the education programme of the public, an audio-visual system has to be used and the attitudes towards consanguineous marriages must change. In premarital screening programme carried on by the government, the couples must be tested and before taking blood they must be informed why the test is done and the genetic counsellor must be sure of the comprehension. It has been shown that a low percentage of at risk couples opted for prenatal diagnosis[9,10]. Experience has shown that even the parents of ß-thalassemia patients have not comprehended the risks and the mental and the financial burden of having a diseased child and continue the pregnancy

without prenatal diagnosis.

In Mediterranean countries as Greece, North and South Cyprus, Italy and in Canada where ß-thalassemia were major health problems, the control programme which had started in 1980's have resulted almost in nil homozygous births by 1990's[11-13,15,16]. The success of the programme was due to a well planned extensive education with large participation along with carrier screening. At the beginning, the target population included doctors, health personel, community and religious leaders and school directors. Above all these, the participation of the government was obtained. Then these informed people extended their knowledge to the public.

The target groups changed from patient families to school leavers, to students, to premarital couples. Scriver's group obtained success in Canada with an organization as cited above and through education of high school students aged 14-18 year along with voluntary blood testing[11].

In conclusion, this experience shows that if we have a large scale education programme encompassing the doctors, the health personel, the community and religious leaders and the government support we will have nil SCA and ß-thalassemia homozygous births.

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