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# Tests for the Prevention of Thalassemia Major in the Couples Referring to Behbahan Health Center, Iran During 2006-2016

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## ABSTRACT

**Background:** Thalassemia is the most prevalent genetic disorder in humans. It is caused by the deficient or lack of production of one of the globin chains. The present study aimed to evaluate the performance indicators of Behbahan Health Center, Iran in terms of the prevention of thalassemia major during 2006-2016.

**Methods:** This descriptive, cross-sectional study was conducted on the couples referring to the pre-marriage counseling centers in Behbahan, Iran for premarital tests during 2006-2016. The participants were selected via census sampling.

**Results:** In total, 24,992 couples were screened, 0.33% of whom (n = 84) were suspected of thalassemia. Out of 84 couples, two cases declared marriage cancellation after electrophoresis. The results of the PND1 test indicated that in 96% of the couples (n = 73), the boy and girl both had thalassemia minor, while in 1.3% of the cases (n = 1), one of them had thalassemia minor, and in 6.2% (n = 2), they were both normal.

**Conclusion:** According to the results, 84 couples were carriers of thalassemia during 2006-2016. Eventually, all the cases with thalassemia major were aborted and prevented from birth.

## 1. Introduction

Thalassemia is the most prevalent genetic disorder in humans. It is caused by the deficient or lack of production of one of the globin chains, which are classified as the alpha, beta, and delta chains [1]. Thalassemia is categorized as alpha thalassemia and beta thalassemia. Beta thalassemia is classified as thalassemia major (severe thalassemia) and thalassemia minor (mild thalassemia). Individuals with thalassemia minor have no problematic anemia. However, if two individuals with thalassemia minor are married, 25% of

their children are expected to have thalassemia major, while 25% of their children will be normal, and 50% will have thalassemia minor [2-4].

Thalassemia occurs in various regions and races across the world, while it has been reported to be more prevalent in the Mediterranean coast, Middle East, and South-East Asia. According to statistics, 15-20% of the inhabitants of regions such as Greece, Italy, South Russia, and India are carriers of the thalassemia gene [5]. Furthermore, approximately, 7% of the world's population has been reported to have thalassemia [6].

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In Iran, approximately 3.5 million individuals are carriers of the thalassemia gene. Beta thalassemia is the most common type of the disease in Iran, which could be classified as mild (thalassemia minor), moderate (thalassemia intermedia), and severe (thalassemia major).

The prevalence of the thalassemia gene in Iran is within the range of 2.5-15%, while this rate has been estimated at 10% in the Caspian Sea and Persian Gulf coastlines and 4-8% in other regions [7]. The most effective strategy to encounter thalassemia will be available when the general knowledge of the disease reaches a level where couples opt for proper preventive behaviors, especially the couples who are the carriers of the disease. Prevention of thalassemia aims at the avoidance of childbirth and contracting severe thalassemia in neonates [8]. One of the key strategies for the prevention of thalassemia major is the screening of marriage applicants.

Since the birth of an infant with thalassemia major depends on the marriage of a man and a woman who are both healthy carriers (thalassemia minor), the marriage may not occur. On the other hand, if the couple deliberately intends to prevent the birth of an infant with thalassemia major, the infant will not be born, and the incidence of the disease decreases. Accordingly, a law was passed in the Islamic Consultative Assembly in 1997, based on which all official registration offices are required to ask marriage applicants to receive thalassemia tests before signing the official marriage certificates [7]. Therefore, the couples referring to official registration offices to conclude their marriage contract must first submit the thalassemia test results in addition to other documents. These individuals often receive a recommendation from the marriage registry and are introduced to laboratories for thalassemia tests [7]. Considering the hereditary nature of the disease, the most effective approach for the parents who are both carriers of thalassemia genes is to receive prenatal diagnostic tests for thalassemia (PND), which are performed at various stages [9]. However, previous studies have indicated that although the incidence of thalassemia major has decreased significantly with the implementation of thalassemia control and prevention programs across Iran, proper planning to enhance the quality of counseling and care services and effective education and information to the target groups in the community are essential to the eradication of thalassemia [10, 11].

Considering that most of the studies conducted in Iran have addressed the prevalence and causes of thalassemia, and the performance of urban health centers for the reduction of the disease has not been thoroughly evaluated, the present study aimed was to investigate the result of tests for the Prevention of Thalassemia Major in the Couples Referring to Behbahan Health Center During 2006-2016.

## 2. Materials and Methods

This descriptive, cross-sectional study was conducted on the couples referring to the pre-marriage counseling centers in Behbahan, Iran to receive premarital tests during 2006-2016. The couples were selected via census sampling.

The required data were extracted from the available records at the Thalassemia Prevention Center in Behbahan, including the data on the total number of the couples (with and without thalassemia), couples with thalassemia minor, couples suspected of thalassemia, couples receiving initial PND tests, couples undergoing the second phase of fetal PND tests, newborns with thalassemia major, and abortions. Data collection was performed using checklists.

Once the data were collected and sorted, they were entered to the SPSS files and analyzed using the following descriptive statistic such as: mean, standard deviation and frequency.

## 3. Results and Discussion

During 2006-2016, 24,992 marriage applicants were screened at the pre-marriage counseling center in Behbahan, Iran. Mean age of the male and female subjects was  $27.38 \pm 5.58$  and  $23.93 \pm 5.55$  years, respectively. Regarding the employment status, the majority of the male applicants were self-employed (50.6%;  $n = 45$ ), while the majority of the women were housewives (71.9%;  $n = 64$ ). In terms of the education level, the majority of the male (34.8%;  $n = 31$ ) and female subjects (37.1%;  $n = 33$ ) had high school diploma. In total, 84 couples (0.33%) were suspected of thalassemia. The total number of the couples, suspected couples, and the results of the electrophoresis test are presented in Table 1.

The highest number of the cases with thalassemia was 14 couples (0.5%) in 2007, as opposed to only two couples (0.07%) in 2009. In addition, two out of 84 couples refused marriage after the electrophoresis test, including one couple in 2011 and one couple in 2016 (Table 1). Among the studied cases, 84 couples were suspected of thalassemia.

The number of the couples receiving PND1 and their test results during 2006-2016 are shown in Table 2. According to the findings, 90.5% of the suspected couples ( $n = 76$ ) received the PND1 test, and 9.5% ( $n = 8$ ) of those receiving electrophoresis did not receive PAND1 due to transient or lack of follow-up. The results of the PND1 test indicated that in 96% of the couples ( $n = 73$ ), both partners had thalassemia minor, while one partner in 1.3% of the couples ( $n = 1$ ) had thalassemia minor, and 2.6% of the cases ( $n = 2$ ) were both healthy. Among 73 couples who were diagnosed with thalassemia minor in the PND1 test, two cases (2.73%) refused marriage. The number of the couples receiving PAND2 and their test results during 2006-2016 are presented in Table 2.

A total of 74 couples (86.9%) who eventually married were taken care of. The follow-up revealed that 9 couples (12.33%) were not pregnant yet. Also, 3 couples (4.1%) had infertility. 1 couple (1.4%) had transient and 1 (1.4%) was not yet been married. Finally, 60 pregnant women (82%) underwent the pnd2 test. Of these, 7 pregnant women (11.7%) were diagnosed with thalassemia major fetuses, all leading to abortion. Ultimately, 30 (50%) of the pregnant women were diagnosed with thalassemia minor fetuses. 23 (38.3%) of the pregnant women were also diagnosed with healthy fetuses (Table 3).

**Table 1:** Total number of couples, suspected couples to thalassemia, and electrophoresis test results in the centers affiliated to Behbahan University of Medical Sciences during 2006-2016

Year	Total number of couples	Number of couples suspected to thalassemia (%)	Number of suspected couples to thalassemia who underwent electrophoretic test (%)	Number of electrophoretic results		Number of cancellation of marriage after electrophoresis (%)
				Minor (%)	Suspected (%)	
2006	2300	5(0.21)	5 (100)	5 (100)		
2007	2780	14 (0.5)	14 (100)	14 (100)		
2008	2500	5 (0.2)	5 (100)	5 (100)		
2009	2700	2 (0.07)	2 (100)	2 (100)		
2010	2465	4 (0.16)	4 (100)	4 (100)		
2011	2470	8 (0.32)	8 (100)	8 (100)		1 (12.5)
2012	2275	11 (0.48)	11 (100)	11 (100)		
2013	2150	7 (0.30)	7 (100)	7 (100)		
2014	1870	13 (0.69)	13 (100)	13 (100)		
2015	1745	4 (0.24)	4 (100)	4 (100)		
2016	1737	11 (0.63)	11 (100)	11 (100)		1 (9)

**Table 2:** Results of pnd1 test for couples suspected to thalassemia in the centers affiliated to Behbahan University of Medical Sciences during the years 2006-2016

Year	Number of couples suspected to thalassemia (%)	Number of suspected couples who underwent the PND1 test (%)	Number of both partners are thalassemia minor (%)	PND1 result		Number of healthy (%)	Number of cancellation of marriage after the result (%)
				Number of one partner is thalassemia minor (%)			
2006	5 (0.21)	5 (100)	5 (100)				
2007	14 (0.5)	13 (92.8)	12 (92.3)	1 (7.7)			
2008	5 (0.2)	4 (80)	3 (75)			1 (25)	
2009	2 (0.07)	2 (100)	2 (100)				
2010	4 (0.16)	4 (100)	4 (100)				
2011	8 (0.32)	7 (87.5)	7 (87.5)				
2012	11 (0.48)	11 (100)	11 (100)				
2013	7 (0.30)	7 (100)	6 (85.7)			1 (14.3)	
2014	13 (0.69)	10 (76.9)	10 (100)				
2015	4 (0.24)	3 (75)	3 (100)				
2016	11(0.63)	10 (90.9)	10 (100)				1 (10)
<b>Total</b>	<b>84(2.7)</b>	<b>76 (90.5)</b>	<b>73 (96)</b>	<b>1 (1.3)</b>		<b>2 (2.6)</b>	<b>2 (5.2)</b>

Within the past two decades, like the other regions in Iran, Behbahan has participated in the national plans for thalassemia prevention, most notably premarital diagnosis. Investigation of all the screened couples in Behbahan city during 2006-2016 indicated that 84 couples were carriers of thalassemia. Eventually, all the cases with thalassemia major were aborted and prevented from childbirth. Our findings also showed that a high percentage of thalassemia minor carriers carried on with their high-risk marriage, so that only two cases (2.73%) among these couples refused to marry after the PND test.

In Sistan and Baluchestan province (Iran), the rate of marriage refusal among the carriers of thalassemia minor was reported to be 3.5% in 2000 and 0.5% in 2002 [12]. The results obtained by Jafari et al. (2007) in Gorgan province (Iran) (2006) were consistent with the present study in terms of the rate of withdrawal from marriage [7]. The observed performance in the present study was poorer compared to most of the studies conducted in Iran, so that the rate of withdrawal from marriage among the carriers of thalassemia minor in Fars province was reported to be 62.1% in 2000 and 48% in 2007 [12]. In the study by Kosarian et al. (2003) on thalassemia minor couples in Mazandaran province (Iran) during 1992-2002, the mean rate of marriage refusal was 51% over the whole years, which denoted increased awareness regarding the disease, which was a significantly higher rate compared to the current research [13]. In the study by Jafari

et al. (2007), 107 thalassemia carrier couples were identified during 1997-2003, who were advised to refuse the marriage; however, only 43% of the couples refused marriage, which was a higher rate compared to the results of the present study [7]. In the present study, 90.5% of the couples suspected of thalassemia (n = 76) received the PND1 test and only 9.5% (n = 8) of those undergoing electrophoresis received the PND1 test due to transient or lack of follow-up. According to the findings of Rezaie et al. (2016) only 11.7% of the 273 couples carrying the thalassemia gene in Sistan region (Iran) had knowledge regarding the disease, while 48.9% underwent the first stage of the PND test, and 38.7% underwent the second phase, which was a lower rate compared to the present study [5]. Recently, the need to strengthen the genetic counseling teams in special counseling centers and their continuous education have been emphasized, as well as the follow-up of the suspected couples and carriers of the thalassemia gene. Furthermore, educating individuals regarding the prevention of thalassemia at the pre-marital age, especially in schoolchildren, plays a pivotal role in the success rate of the thalassemia prevention plan [14].

The follow-up carrier couples and their referral to health centers after undergoing the first stage of the PND test is of paramount importance since after the genetic test, there is no  $\beta$ -globulin gene mutation in most of the cases. Naturally, these cases are eliminated from care coverage in order to reduce the costs imposed on the healthcare system [15].

**Table 3:** Results of pnd2 test in the centers affiliated to Behbahan University of Medical Sciences during 2006-2016

Year	Couples suspected to thalassemia (%)	Number of pregnant women who underwent the PND2 test (%)	PND2 result			Number of abortions (%)
			Thalassemia major (%)	Thalassemia minor (%)	Healthy (%)	
2006	5 (0.21)	5 (100)	3 (60)	1 (20)	1 (20)	3 (60)
2007	14 (0.5)	12 (85.7)		(25)	9 (75)	
2008	5 (0.2)	2 (40)		2 (100)		
2009	2 (0.07)	2 (100)		1 (50)	1 (50)	
2010	4 (0.16)	4 (100)	1 (25)	1 (25)	2 (50)	1 (25)
2011	8 (0.32)	4 (50)		2 (50)	2 (50)	
2012	11 (0.48)	11 (100)	1 (9)	7 (64)	3 (27)	1 (9)
2013	7 (0.30)	4 (57)	1 (25)	2 (50)	1 (25)	1 (25)
2014	13 (0.69)	6 (4.6)	1 (16.7)	4 (66.6)	1 (16.7)	1 (16.7)
2015	4(0.24)	2 (50)		1 (50)	1 (50)	
2016	11 (0.63)	8 (73)		(75)	2 (25)	
<b>Total</b>		60 (75)	7 (11.7)	30 (50)	23 (38.3)	7 (11.7)

Considering the normal family size in Iran (four members, including two children per each couple) and the risk of giving birth to a neonate with thalassemia major per each minor couple (25%), if these tests could prevent the marriage of thalassemia carriers, 42.5% of the births of the children with the disease could be prevented within an 11-year period in the present study (2006-2016).

This research is one of the few studies focusing on the performance of urban health centers for the reduction of thalassemia. In addition, the study was conducted in a high-risk region for thalassemia, which could be considered as one of the strengths of the current research. However, the present research was conducted on a city level, and further investigations are required are larger scales in order to provide accurate results for healthcare planners and managers in Iran since the findings may not be fully generalized to the whole country due to the cultural, social, economic, and geographical differences in various provinces.

#### 4. Conclusion

According to the results of premarital tests, 84 couples were carriers of thalassemia during 2006-2016. Eventually, all the cases with thalassemia major were aborted and prevented from childbirth. Therefore, it is necessary to give parents the necessary information.

#### Authors' Contributions

S.M.K., designed the article. T.R. and E.M.Sh., did write the article E.M.S, was supervisor. S.S.H., edited the article. All authors read and approved the final manuscript.

#### Conflict of Interest

The authors declare no conflict of interest.

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