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Pre-marriage Sickle Cell Screening Program in South Region of Iran, A Pilot Study on 50 Cases of Sickle Trait

Khoda Morad Zandian[1], Mohammad Pedram[1], Fatemeh Kianpour Ghahfarokhi[2]

1. Research Center of Thalassemia and Hemoglobinopathies, Ahwaz Jondishapur University of Medical Sciences, Ahwaz, Iran
2. Faculty of Education and Psychology, Shahid Chamran University,Ahwaz, Iran.

Corresponding Author: Khoda Morad Zandian, Assistant Professor of Pediatric Hematology and Oncology, Jondishapour University of Medical Sciences, Ahvaz, Iran. P. O. Box: 6335733119, Tel: +98(611)3743285, Fax: +98(611)3743289, E-mail: drzandian@yahoo.com.

Abstract

Background: Studies have demonstrated that sickle cell trait can be found in an asymptomatic healthy carrier with normal complete blood count (CBC) and red blood cell (RBC) indices. According to Iranian Ministry of Health bulletin instructions, prenuptial Thalassemia Screening Program (TSP) primarily depends on RBC indices which are measured through a routine CBC. Only when these levels are below the standard values in both couples, hemoglobin electrophoresis is performed. Because of normal values of RBC indices in the sickle cell trait, prenuptial TSP might miss it.

Materials and Methods: To approach and prove this hypothesis, we carried out a prospective randomized pilot study of one year duration on 50 known cases of sickle cell trait (the parents of sickle cell and sickle-Thalassemia referral cases to Ahvaz Sickle Cell Center, affiliated to Ahvaz Jondishapour University of Medical Sciences). An informed written consent was obtained from all patients. Complete blood count was done including a blood film, and CBC was done by Helena Automated Cell Counter. Sickle preparation was done by freshly made sodium bisulfate 2%. It was read after one hour for the first time and was then read after 24 hours by macroscopic and microscopic methods. Hemoglobin electrophoresis was done by Helena apparatus. Serum ferritin was measured by Immunoradiometric Assay (IRMA). Iron deficient cases were treated by oral Iron agents.

Results: Data of the first screening line showed MCV>80 and MCH>27 in more than 94% and HbA2 value of less than %3 in %100 of sickle cell trait cases.

Conclusion: Sickle cell trait with normal indices might be skipped through pre-marriage TSP, and their carrier status is not determined without performing hemoglobin electrophoresis. We recommend mandatory hemoglobin electrophoresis and sickle preparation to be added to the first line of prenuptial Thalassemia Screening Program in the sickle target zones such as the southern parts of Iran.

Keywords: sickle cell anemia, diagnosis, Iran

Introduction

Clinical Practice of medicine has shown that the prevalence of sickle cell trait is high in the southern part of Iran, especially in Khuzestan province at the border of Iraq and Persian Gulf.[1,2] Clinical assessment has demonstrated that sickle cell trait is usually asymptomatic with normal CBC and RBC indices. It is a carrier state with hemoglobin A > S and normal blood smears with few target cells and is not associated with anemia.

A sickle trait is not considered as sickle cell disease because it does not cause a clinical illness and the affected person is a healthy carrier. It is the most benign form of the sickling disorders which does not seem to affect life span and the quality of life.[1,2,3] According to Iranian Ministry of Health bulletin instruction (May 1999)[4] prenuptial Thalassemia Screening Program primarily depends

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on RBC indices (MCV, MCH) which are measured through a routine CBC using Helena Electronic Cell Counter. Only when these levels are below the standard values in both couples, hemoglobin electrophoresis is performed.[2,3]

Because of normal values of RBC indices in the sickle cell trait, prenuptial TSP might miss its diagnosis.

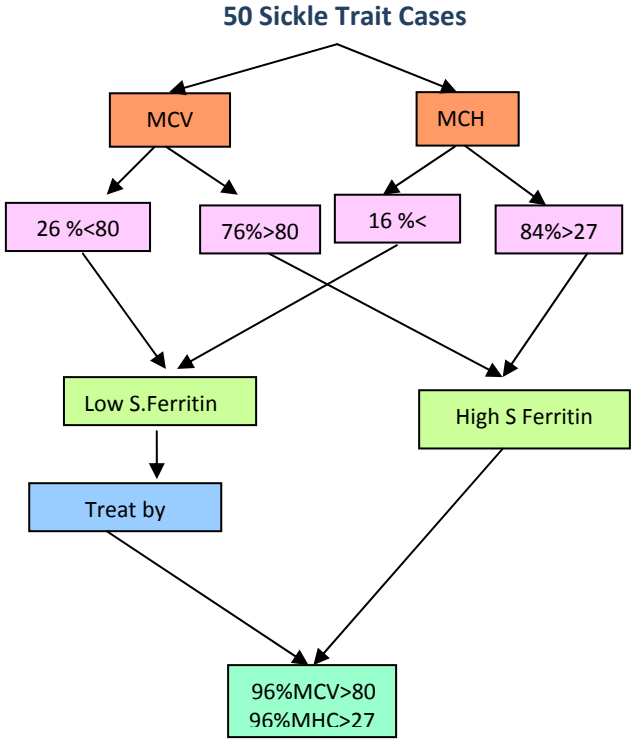


Diagram 1: *First and Second Line of MCV and MCH Percentages*

Materials and Methods

To approach and prove this hypothesis, we carried out a descriptive cross-sectional prospective pilot study of one year duration (Sept.1999-Sept .2000) on 50 known cases of sickle cell trait (the parents of sickle cell or sickle-Thalassemia referral cases to Sickle Cell Center, Ahvaz Shafa Hospital, affiliated to Ahvaz Jondishapur University of Medical Sciences). An informed written consent was obtained from each participant. Complete blood count was done including a blood film, and CBC was done by Helena Automated Cell Counter. Sickle preparation was done by freshly made sodium bisulfate 2%. It was read after one hour for the first time and was then read after 24 hours by macroscopic and microscopic methods. Hemoglobin electrophoresis was done by Helena apparatus. Serum ferritin was measured by Immunoradiometric Assay (IRMA). Iron deficient cases were treated by oral Iron agents.[3]

Results

Data from the first and the second screening lines have shown a normal MCV>80, and MCH>27 in 96% (Figure1) and normal hemoglobin A2 values of less than 3% in 100% of cases. Abnormal hemoglobin S<50% with average (30-46%) was seen in 100% of cases (Table1). Positive sickle preparation was seen in 96% (Table2) and negative sickle preparation in 4% of all randomized cases.

Discussion

Clinical Practice of medicine has shown that the prevalence of sickle cell trait is high in the southern part of Iran, especially in Khuzestan

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province at the border of Iraq and Persian Gulf and Oman-sea.[1,2] Prenuptial Thalassemia Screening program is not sensitive enough to detect sickle cell trait cases so the carriers might be overlooked in the prenuptial Thalassemia Screening Program.[2,5] We recommend mandatory

Table1: *Number of Patients with HbS<50%*

HbS%	No
<30%	4
30-33%	16
34-37%	8
38-40%	7
40-45%	13
44%	1
46%	1
Total	**50**

Table2: *Results of Preparation in Ethnic Randomized Cases*

Cases	NO:	S.C.prep
Arab	44	+
Bakhtiari	2	-
Kurd	1	+
Balooch	1	+
Lengeh	1	+
Busheheri	1	+
Total	**50**	

hemoglobin electrophoresis and sickle preparation to be added to the first line of Prenuptial Thalassemia Screening program in the sickle target zones such as the southern parts of Iran to roll out sickle trait before marriage.[2,6,7] because pregnancy with double compound heterozygote of sickle-Beta Thalassemia disease can be as clinically severe as sickle cell disease depending on the severity of Beta Thalassemia variant. Other same patterns for recessive gene transformation still remain and their carrier statuses are not determined.[1,2,4] Available Thalassemia screening programs are not designed for sickle cell screening. Therefore, we need a new, simple, proven, practical algorithm to approach and tackle these problems. In a simple way (as we did locally), we recommend that High Performance Liquid Chromatography (HPLC) or hemoglobin electrophoresis plus sickle preparation should be added to the first line of the prenuptial Thalassemia Screening Program in sickle prevalent zones in southern parts of Iran.

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