JOURNAL OF CURRENT RESEARCH IN SCIENCE (ISSN 2322-5009)
CODEN (USA): JCRSDJ
2014, Vol. 2, No. 6, pp: 869-872
Available at www.jcrs010.com

REVIEW ARTICLE

THALASSEMIA AS THE MOST PREVALENT BLOOD DISORDER IN IRAN. A REVIEW

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ABSTRACT: thalassemia which is the most prevalent blood disorder in Iran occurs as a result of a genetic disorder in the natural structure of hemoglobin chain. Since Iran is, geographically speaking, located on the thalassemia belt, the prevention and control of this disease is of a great significance. A search was conducted in available scientific databases in Iran such as SID, IranMedex, Medlibe and Google Scholar . The following keywords were used in the searching process: thalassemia, prevention, genetic disorder, beta thalassemia, Hemoglobin. In order to cut down on physical and mental side effects on a patient and his family as well as to reduce the costs of this genetic disease both for patients' families and society, the best solution according to previous research is prevention.

Key words: thalassemia, prevention, genetic disorder

INTRODUCTION

The blood of a healthy human adult contains three hemoglobin, HbA, HbA2 and HbF which respectively comprise 96%, 1-3% and .5-15% of the total body hemoglobin(1, 2).

Thalassemia refers to a group of diseases which appear as a result of a genetic disruption in the natural structure of a hemoglobin chain. This is in fact the most prevalent genetic disease among human beings and is increasingly recognized as a main healthcare threat throughout the world(3-6). It either stops or hinders the production of hemoglobin, and since the main type of hemoglobin after the birth is hemoglobin A, a disruption in the production of each of its chains would lead to alpha or beta thalassemia. The geographical belt of this disease is located in Mediterranean countries, western south of Europe to Asia and a wide area of central Africa. In Iran it is mostly prevalent in the coastlines of the Caspian and Oman Seas. Estimations show that 3% of the world populations are carriers of beta thalassemia gene. In Iran, about three million people are estimated to carry this gene and about twenty thousand people are estimated to be afflicted with major thalassemia (7-9).

In Iran, beta thalassemia is the most prevalent type in either of these forms: minor thalassemia, carrier of intermediate thalassemia and major thalassemia(10).

Considering the fact that Iran is geographically located on the thalassemia belt, diverse research has been conducted so far to determine the percentage of carriers in different regions. According to such research, the percentage of carriers in Sanandaj is estimated to be about 3.5%(11), in Kerman 5.7%(12), in Kashan 3.32%(13) and in Birjand it is reported to be 1%(14). Within a study conducted on 15,974 volunteers in Gorgan, the percentage of carriers was reported to be 2.75% among whom 2.99% were male and 2.5% were female(15).

A myriad of research has been conducted in Iran and other parts of the world to investigate the side effects of major thalassemia(16-24).

Today, in order to grow naturally and continue to live, thalassemic patients use recurrent blood injections(25, 26). Besides its useful effects, this treatment has many hazardous side effects such as infection, iron deposition in different tissues which could lead to heart failure, liver failure, hypo-thyroid, diabetes mellitus(16) and hypoparathyroidism(26-29).

Moreover, similar to other chronic diseases, the physical, social, mental and economic side effects of this disease are so vast. Thalassemia would affect family, healthcare system and society. The

money and time a family should spend on treating this disease as well as the costs imposed on society and its healthcare system for recurrent injections and employment of a medical staff are all the function of this disease. Among the other side effects of this disease are growth disorder, short height, hearing loss, depression and low quality of life(6, 30-34).

Thalassemia is a preventable disease. If today it does not get restricted and simpler with the help of proper scientific interventions, in near future we will all be faced with an acute problem within the thalassemia belt. The oldest prevention method was employed in Italy in 1943 in the thalassemia fighting center. In this method, pre-marriage screenings along with abortion therapy were used as successful methods. In England, these two methods were used in 1977 for Cyprus, Asian and African immigrants(35).

Generally speaking, there exists three preventive methods with this regard including screening the marriage volunteering population, preventing the marriage of two gene carriers, early pre-birth diagnosis, abortion therapy and using genetic engineering to treat a fetus afflicted with thalassemia inside the womb(9, 36).

Considering the high prevalence of this disease, preventive programs are of a great significance today. National attempts at preventing thalassemia have begun since the last two decades in Iran (6, 37, 38). In this country, the preventive program is fundamentally based on pre-marriage screening and consultation(6, 38).

It needs to be mentioned that a lack of awareness and correct perception of thalassemia acts as an obstacle in the way of preventing this disease(39). Besides awareness raising, modifying people's attitudes towards a certain healthcare issue is another primary step to alter their behavior. In a study, it was revealed that face to face instruction can significantly affect people's attitude(40).

Wong et al.(7) found a higher awareness of thalassemia among people of a higher income, education and more professional and managerial occupations(7).

CONCLUSION

According to the previous body of research, prevention is the best policy for reducing physical and mental side effects on both patients and their families as well as the imposed costs on society. Initially we could begin with raising the society's awareness and cognition of thalassemia. Then, preventive methods can be used such as screening the to-be-married population in order to prevent the marriage of the disease carriers. We can also take advantage of pre-birth diagnosis, abortion therapy and genetic engineering to treat an afflicted fetus within the womb. In order for the family adjustment plans to be correctly enacted in Iran's Thalassemia center, first of all the family adjustment status of the couple should be looked into. Its precise descriptors should be determined so that this background information can be used to pave the way for an effective instruction.

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