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THALASSEMIA AWARENESS AMONG HEALTHCARE PROVIDERS

Hala Alsoukhni MD* and Majd Alsoukhni,

*Royal Medical Services, Department of Stem Cell Transplantation, Military Cancer Center, Amman-Jordan.
Global Health Development/ Eastern Mediterranean Public Health Network, Amman-Jordan.

***Corresponding Author: Hala Alsoukhni MD**

Royal Medical Services, Department of Stem Cell Transplantation, Military Cancer Center, Amman-Jordan.

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ABSTRACT

Objective: The aim of this survey is to determine the awareness about thalassemia among healthcare providers in two Royal Medical Services Hospitals. **Methods**: This cross-sectional study was conducted among healthcare professionals randomly selected in Prince Rashid Hospital in Irbid and Prince Hashim Hospital in Zarqa in Jordan. The employee who participated in this survey include physicians from different specialties, dentists, pharmacists, nurses, and laboratory technologists. The questionnaire contained 14 close ended questions regarding facts about thalassemia, disease transmission, parents' consanguinity, screening, prevention and treatment. **Results**: Among 100 healthcare provider 64 were males and 36 were females. Fifty were physicians, 3 were dentists, 24 were laboratory technologist, 18 were nurses, and 5 were pharmacists. Responses to questions were 100%. Around 80% of responses were right.

Conclusion: The awareness about thalassemia among healthcare professionals is adequate however; more education is necessary for this category about the disease transmission, prevention, and role of carriers' recognition so they can be involved in a national programs for public awareness about thalassemia to help in disease eradication in Jordan.

KEYWORDS: Thalassemia, Awareness, Survey.

INTRODUCTION

Thalassemia is a group of hereditary blood diseases characterized by quantitative defect of globin chain production which cause ineffective erythropoiesis or hemolysis and anemia. Hemoglobin has heme group part and protein part which is globin chain. Normally in adults the major hemoglobin is Hemoglobin A which consists of 2 identical α-chains and 2 identical β-chains, each chain is linked with one heme group.[1] The main two types of thalassemia are α and β thalassemia. α-thalassemia results from gene deletion, there are 2 α-globin gene on each chromosome 16, so 4 genes are required to make α-globin chains. If one or two genes are deleted the result is α-thalassemia minor. If more than 2 genes are deleted, the result is α-thalassemia major with moderate to severe anemia that occur due to excess β-chain that causes hemolysis. Hemoglobin H disease results when 3 genes are deleted, and hemoglobin Bart's (Hydrops Fetalis) result when the baby born with complete absence of α-genes which is incompatible with life. There are 2 genes for β-globin chains on chromosome 11. B-thalassemia results mostly from single base substitution that produce defective unstable β-chains. If one of the two genes are affected, the result is β-thalassemia minor, if the two genes are affected the result is β-thalassemia major (Cooley's anemia) with markedly decreased or absent production of β-chains. This result in excess α-chains which are extremely unstable and cause ineffective erythropoiesis and severe anemia.[2] Clinical signs and symptoms include symptoms of anemia, jaundice, enlarged spleen, facial bone deformities, growth retardation, delayed puberty, and complications of iron overload due to frequent blood transfusion.[3]

The inheritance of thalassemia is autosomal recessive which means that both parent should carry the trait to transmit the disease to their offspring. Thalassemia is common in Middle East, East Asia, Mediterranean and African countries.[4] The prevalence in these regions may reach as high as 10%.[5]

Management of thalassemia major depends on regular blood hypertransfusion to maintain hemoglobin level more than 9.5 g/dL. Iron chelation to prevent iron overload. The patients need multidisciplinary team including hematologist, endocrinologist, cardiologist, hepatologist, psychologist, genetic counselor, and social worker. Life expectancy depends on blood transfusion and iron chelation, with optimal conditions it is around 40 years.[6] The only curative treatment for thalassemia are allogenic stem cell transplantation and gene therapy.[7]

Prevention of thalassemia is the most important program in thalassemia management, it includes carriers screening, premarital counseling and prenatal diagnosis. The program should focus on public awareness and education.[8]

In Jordan, consanguineous marriage is quite common with consanguinity rate exceed 28.5%, but in June 2004 the premarital screening and counseling became mandatory by law to decrease carriers' marriage.[9] Also antenatal diagnosis is available for suspected cases but abortion is illegal in Jordan. So, the only way to decrease prevalence of thalassemia is through educating the population about this disease. Intensive education of healthcare providers is needed to help in public education.

The aim of this study is to evaluate the awareness of thalassemia among healthcare providers.

MATERIALS AND METHODS

One hundred questionnaires were answered by different fields' healthcare providers in two military hospitals in Jordan (Prince Rashid Hospital in Irbid and Prince Hashim Hospital in Zarqa) over two months July and August 2020. The participants were chosen randomly from different departments, different ranks and genders. The questionnaire comprises fourteen close ended questions about etiology of thalassemia, the role of consanguineous marriage in transmission of the disease, sample needed for diagnosis, if the disease is preventable and curable, the role of premarital counselling in prevention of thalassemia and is there any other way for prevention, detection during pregnancy, the role of carrier screening in prevention of the disease, thalassemia minor, the possibility of disease transmission between carriers, and the need for blood transfusion.

The collected data were reviewed and statistically analyzed using SPSS program.

1-Is thalassemia a genetic disease?	Yes	No
2-Do consanguineous marriages have any role in incidence of thalassemia?	Yes	No
3-Can thalassemia be diagnosed by blood test?	Yes	No
4-Is thalassemia a preventable disease?	Yes	No
5-Is thalassemia a curable disease?	Yes	No
6-Is premarital consultation useful for prevention of thalassemia?	Yes	No
7-Is premarital consultation is the only way to prevent thalassemia?	Yes	No
8-Can thalassemia major be diagnoses prenatally?	Yes	No
9-Does recognition of thalassemia minor have any role in prevention of thalassemia major?	Yes	No
10-Does thalassemia minor have any signs or symptoms?	Yes	No
11-Is thalassemia minor curable?	Yes	No
12-If two persons with thalassemia minor get married, is there any chance to have children with thalassemia major? Yes No		
13-Does marriage between a normal person and person with thalassemia minor can result in thalassemia major? Yes No		
14-Do thalassemia major patients need blood transfusion for the whole life?	Yes	No

RESULTS

Sixty four of the participants in this survey were males and 36 were females. Fifty were physicians, 3 were dentists, 24 were laboratory technologist, 18 were nurses, and 5 were pharmacists. All of them responded to all questions. All of the participants (100%) were aware of the etiology and inheritance of thalassemia, 95% were aware about the role of consanguineous marriage in incidence of thalassemia, 96% knew that thalassemia is diagnosed by blood test, 74% of them knew it is a preventable disease, 85% knew that thalassemia is incurable, 95% were aware about the role of premarital consultation in thalassemia prevention, only 41% have the knowledge that there is other method for thalassemia prevention beside premarital consultation, 85% knew that thalassemia can be diagnosed prenatally, 78% were aware of the role of thalassemia minor recognition in prevention of thalassemia major, 37% said that thalassemia minor patients have symptoms, and 82% said it is incurable, 90% recognized that marriage between two persons with thalassemia minor may result in thalassemia major children, 14% said that marriage between normal person and a person with thalassemia minor may produce children with thalassemia major, 85% knew that thalassemia major need blood transfusion for the whole life.

DISCUSSION

The level of knowledge about thalassemia in this study is quite good (around 80%). A survey conducted in Kolkata Hospital in India among junior doctors revealed that 77.7% of the participants have adequate knowledge about thalassemia which is close to our result.[10] Another survey conducted in Delhi over medical students showed slightly lower level of knowledge (75%).[11] In 2015 Haque et al. carried out a survey among the students of Royal college of medicine in University of Kuala Lumpur, Malaysia to determine the level of awareness on thalassemia and the results showed a very good level of knowledge (90%).[12]

Our participants have a very good knowledge about the etiology of thalassemia as all of them answered that it is genetic disease, and the majority (95%) acknowledged

the role of consanguineous marriage in disease transmission and the importance of premarital consultation in disease prevention but less percentage (78%) recognized the role of thalassemia trait recognition in prevention of thalassemia major which means some lack in knowledge about mode of inheritance of the disease among participants. Rabbani et al. performed a survey on future healthcare providers in Ras Al Khaimah University in UAE and reported a high level of thalassemia awareness among students from Middle East which is attributed to the fact that thalassemia is common in the Middle East countries.[13]

A quiet high percentage of participants (59%) were not aware about prenatal diagnosis as a preventive method for thalassemia major, this could be ascribed to the fact that abortion is illegal in Jordan.

This study have some limitations. The sample size is small and is not fair to compare between different professions responses.

CONCLUSION

Awareness about thalassemia among healthcare professionals is adequate however; more education is necessary for this category about the disease transmission, prevention, and role of carriers' recognition in prevention, so they can be involved in a national programs for public awareness about thalassemia to help in disease eradication in Jordan.

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