INTRODUCTION

Thalassemia are heterogeneous group of hereditary disorders in which there is a reduced rate of synthesis or production of one or more of the globins’ polypeptide chains which leads to ineffective erythropoiesis and anaemia. Normal hemoglobin, also called hemoglobin A, has four protein chains—two alpha globin and two beta globin. The two major types of thalassemia, alpha and beta, are named after defects in these protein chains. Four genes (two from each parent) are needed to make enough alpha globin protein chains. Alpha thalassemia trait occurs if one or two of these four genes are missing. If more than two genes are missing, severe anemia occurs. The most severe form of alpha thalassemia is called alpha thalassemia major or hydrops fetalis. Babies who have this disorder usually die before or shortly after birth. Two genes (one from each parent) are needed to make enough beta globin protein chains. Beta thalassemia occurs if one or both genes are altered.

The severity of beta thalassemia depends on how much one or both genes are affected. If both genes are affected, the result is moderate to severe anemia. The severe form of beta thalassemia is known as thalassemia major or Cooley's anemia. Current approaches include hematopoietic stem cell transplantation. Disease management includes prenatal diagnosis, transfusion therapy, bone marrow transplantation, and out of these therapy only bone marrow transplantation (BMT) is potentially curative for thalassaemia patients [1].

The thalassemia can be classified according to which chain of the Hb hemoglobin molecule is affected. There are two main types of thalassemia.

- Alpha thalassemia occurs when a genes or genes related to alpha globin protein are changed (mutated) or missing.
- Beta thalassemia mostly occurs when similar gene defects affect the production of the beta globin protein. [2].

Signs and Symptoms of thalassemia include severe tissue hypoxia, Iron overload, Hemolytic anemia, Shortness of breath, Bone deformities, Dark urine, Slow growth, delayed puberty, Jaundice and An enlarged spleen, heart and liver.

Thalassemia is a hereditary anaemia of varying severity is one of the common inherited disorders in Pakistan [3]. It is estimated that about 9000 children with beta thalassemia are born in every year, although no documentary registry is available in Pakistan. The estimated carrier rate is 5-7%, which accounts to 9.8 million carriers in the total population [4]. The cultural and religious scenario in Pakistan is such that consanguineous marriages are quite common and there is no concept of premarital screening or counseling of individuals with a family history of the disease. Furthermore there is no antenatal diagnosis widely available.

The only way to prevent the disease and reduce the mortality is by educating the general Population[5]. For this reason in this present study, awareness among general population, parents of thalassemia patients, student of Pharm D, MBBS regarding the disease was evaluated. Same as awareness about HPV vaccine have been studied by our research group [6]. These types are studies are useful for medical professionals.

MATERIALS AND METHODS

Questions regarding the duration of illness, family history of thalassemia, consanguinity of parents, screening of blood, knowledge about the infections transmitted through blood products and prevention from these infections, mode of transmission of disease, awareness regarding iron chelation therapy and antenatal diagnosis were asked. All the data was entered in SPSS version 20 and evaluated. Results have been presented in the form of frequencies and percentages where applicable.

RESULT

We’ve determined the awareness ratio and found out that the awareness rate of thalassemia was not up to the mark as we thought. Among 200 population...
only 22% of the people had good knowledge about thalassemia and rest 78% of people were not well aware of the disease as well as its consequences. By using SPSS software we apply chi square p values between male & female highly significant indicate no awareness of thalassemia. Also the result of different population have highly significant result.

![Figure 1: Awareness of Thalassemia in Pharm D, MBBS and other population (male, female)](image)

In our survey of thalassemia that we did we observed different aspects of this disease. Thalassemia is basically a life threatening hereditary disease which is caused when two minor thalassemia known as “carriers” mate to produce a child which results in a major carrier. One in 4 children are affected by this incurable disease but we can maximize the survival for some years of life by transfusion of blood once in every 15 days or a month as required by the patient. Practically it is not possible to keep transfusing the blood due to less availability of blood in blood banks and also as no. of patients is increasing every year. We can also perform the bone marrow transplantation (BMT) to save life but it is unpredictable as the patient may accept or reject the transfused bone marrow.

Our results show that there is a minimal ratio of awareness regarding thalassemia in common population as compared to medical students. This survey was conducted among the Pharmacy students' of Jinnah University for women and among the students of MBBS from Sindh Medical University, and third survey was done in the random population. Awareness programs such as seminars, workshops, walks, health care lectures, pamphlets, marathons, advertisements on print and electronic media for donation of blood and counseling the general public to avoid marriages where both are minor carriers should be conducted. Concrete step must be taken to test the couple before they head for a family so that the thalassemia rate is minimized which presently is 1 crore 5 lac. If such serious measures would be taken thalassemia would be eradicated within 5 to 10 years INSHALLAH!

REFERENCES