

Analysis of Profile of Blood Transfusion in Patients of Beta Thalassemia: An Experience from Tertiary Care Centre of Jammu Region

Dr. Vijay Sawhney¹, Dr. Salve Sharma²

¹Ex Professor and Head, Department of Transfusion Medicine, GMC Jammu, India

²GMC Jammu, India

Abstract: *Thalassemia is an autosomal recessive blood disorder in which there is reduced rate of synthesis of one of the globin chains that make up the hemoglobin. Ineffective bone marrow erythropoiesis and excessive red blood cell hemolysis together account for the anemia. This study is conducted at a tertiary care hospital and found that 255 (92.39%) patients were diagnosed as Beta thalassemia major, 19 (6.88%) patients were diagnosed as beta thalassemia intermedia and 2 (0.72%) patients suffered from beta thalassemia minor. Out of registered beta thalassemia cases, 243 patients received regular blood transfusion therapy in the form of packed red cells. District-wise distribution of thalassaemic patients was observed and found that majority of patients belonged to Jammu and Rajouri districts. Family history of thalassemia was found in 79 (28.62%) of patients. Thalassemia patients comprise the group receiving regular transfusion. Good infrastructure and quality care to these patients is of prime importance. Policies need to be reviewed to provide maximum benefit to these patients.*

Keywords: Thalassemia, Blood transfusion, packed red cells

1. Introduction

Thalassemia is an autosomal recessive blood disorder in which there is reduced rate of synthesis of one of the globin chains that make up the hemoglobin. Ineffective bone marrow erythropoiesis and excessive red blood cell hemolysis together account for the anemia. Beta-thalassemia is caused by the reduced (beta) or absent (beta) synthesis of the beta globin chains of the hemoglobin tetramer. Three clinical and hematological conditions of increasing severity are recognized, i.e., the beta-thalassemia carrier state, thalassemia intermedia, and thalassemia major. (Cao A et al. 2010). According to a 2008 report from the World Health Organization, more than 40 000 infants are born with β -thalassemia each year, of whom about 25,500 have transfusion-dependent β -thalassemia. (Modell B et al. 2008). The thalassemsias are among the most common genetic disorders worldwide, occurring more frequently in the Mediterranean region, the Indian subcontinent, South east Asia, and West Africa.

It has been generally estimated that, every year 50 thousand to 1 lack children, living in the low and middle income countries die because of this chronic genetic ailment (beta thalassemia major), however, an estimated population of 7% in the entire world is the carrier of hemoglobin disorder in the form of beta thalassemia major (Arif et al. 2008). Studies show the propagation of beta thalassemia major to be a serious threat to the middle- and low-income countries across the world (Khalid et al. 2019).

Blood transfusion therapy is the mainstay of treatment in Beta thalassemia patients. Around 100000 children are born every year with severe homozygous state of the disease in India. The objective of this study was to assess clinical data and transfusion profile of beta thalassemia patients attending tertiary care hospital. . It is well recognized that even in geographic regions where b-thalassaemia is common, not all ethnic groups are at the same risk of possessing the

thalassaemia gene. In India, where the average prevalence of the beta thalassaemia trait is about 3.5 %, Sindhis and Punjabis are known to carry the b-thalassaemia gene more commonly than other Indian populations. (Madan N et al. 2010)

2. Methods and materials

This prospective study was done at a tertiary care hospital SMGS Hospital, Government Medical College, Jammu from January 2019 to December 2019. Beta thalassemia patients who received transfusion therapy in the form of packed red cells were included in the study. A Proforma was prepared which included the following parameters: age of diagnosis, age group of the presenting patients, sex, religion, districts of the state they belonged to, family history of thalassemia, blood group, type of thalassemia major/intermediate/minor, total no. of transfusion received, chelation therapy status. . These parameters were obtained from patients previous records at department of transfusion medicine and pediatrics. Patient consent and hospital ethical committee clearance was taken. The diagnosis had been previously established based on blood film, electrophoresis and hemoglobin F quantitation performed at various other centres and departments. The blood component details and data was analysed. The patients were followed post transfusion for the development of adverse transfusion reactions.

3. Result

Total registered cases of Thalassemia in our centre are 276. Out of them, 156 (56.52%) were males and 109 (39.49%) were females. Based on types of Thalassemia, 255 (92.39%) patients were diagnosed as Beta thalassemia major, 19 (6.88%) patients were diagnosed as beta thalassemia intermedia and 2 (0.72%) patients suffered from beta thalassemia minor. 67 (24.27%) patients were between 10 years to 20 years, 58 (21.01%) patients were older than 20

Volume 10 Issue 7, July 2021

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years and 151 (54.71%) patients were less than 10 years. Out of registered beta thalassemia cases, 243 patients received regular blood transfusion therapy in the form of packed red cells.

Type of Thalassemia	Number of Thalassemics
Thalassemia major	255
Thalassemia minor	2
Thalassemia intermediate	19
Total	276

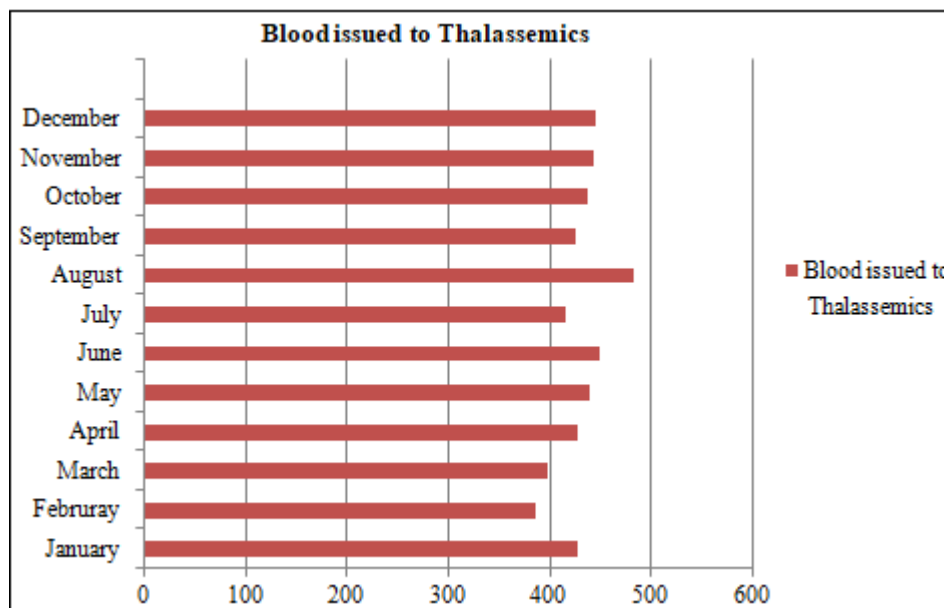
Average number of transfusions per patient in patients less than 10 years was found to be 2 times per month. In patients between 10 years to 20 years, average number of transfusions per patient was found to be 2 to 3 times in a month. In patients more than 20 years, average number of transfusion per patient was found to be 4 times in a month.

District-wise distribution of thalassemic patients was observed. Majority of patients belonged to Jammu (n=98) and Rajouri (n=64) region.

District	Number of Thalassemics
Jammu	98
Rajouri	64
Reasi	24

Poonch	19
Kathua	16
Udhampur	11
Doda	11
Samba	9
Kishtwar	14
Other state	10

Family history of thalassemia was found in 79 (28.62%) of patients. Majority of patients who gave a family history were Muslim (41/88, 46.59%) followed by Hindu (49/156, 31.41%), Sikh (3/21, 14.28%) and Christian (0/11, 0%). Majority of thalassemics were Rh positive 248 and 28 were Rh negative. Most common blood group in thalassemic patients was found to be B positive (n=89) followed by O positive (n=84), A positive (n=50) and AB positive (n=25). Among Rh negative group, O negative (n=10), B negative (n=9), A negative (n=5) and AB negative (n=4). 90.94% (n=251) of beta thalasesemics were on chelation therapy. During the study period, 12420 packed red cells are collected and 5180 units of packed cells were issued to thalassemic patients. Month wise issuance of blood component to thalassemics is shown in Table 1.



4. Discussion

In our study, majority (92.39%) patients were beta thalassemia major followed by thalassemia intermediate and thalassemia minor. Beta thalassemia major patients were completely dependent on blood component transfusion and seek medical care than thalassemia intermediate and thalassemia minor. Talsania et al. (18) in their study in Gujrat found the similar differences in profile of beta thalassemia major , intermediate and minor patients.

Most of the thalassemic patients belonged to Jammu district of the state, this may be due to the fact that hospital here is tertiary care hospital with good facilities and families of thalassemic patients have migrated to Jammu for receiving timely medical care. Attempts should be made to improve treatment facilities including availability of safe blood

component, chelation therapy and transfusion specialist at district level hospitals so that these families don't have to migrate to cities.

Family history of thalassemia was more common in Muslim population. This can be explained that there is consanguineous marriages in muslims which cause the increase in genetic diseases in this group. The prevalence of beta thalassemia major is especially high in countries where there are close family marriages. (Ghosh S et al. 2008). Genetic counseling will reduce the prevalence of genetic abnormalities in this population. It has also been intensively studied that cousin marriages are the major reasons of beta thalassemia major (Ayub et al. 2017; Faizan-ul-Haq et al. 2016).

Blood transfusion sustains life in beta thalassemic patients but it also comes along with risk of adverse effects of transfusion like transfusion transmitted infections, alloimmunization, transfusion reactions and iron overload. In our study, it was found that majority of beta thalassemics were on iron chelation therapy. Desferrioxamine are being given to treat iron overload. Effectiveness of iron chelation therapy is not monitored periodically due to non availability of test for serum ferritin in the hospital. It is also due to the fact that family of thalassemics cannot afford the test to be done outside the hospital. Lack of social support has also been seen to be a significant factor, which increases psychosocial burden of the disease (Palanisamy et al. 2017).

Thalassemia patients comprise the group receiving regular transfusion. Good infrastructure and quality care to these patients is of prime importance. Policies need to be reviewed to provide maximum benefit to these patients.

As there is huge financial burden on the families of thalassemics patients, it is suggested to come together as a community for their help. Social workers, local governing bodies and corporate sector need to work together. In this regard, it is suggested that one child should be adopted to provide that child the best possible medical care.

As patients from far flung areas have to reach to tertiary hospital in the cities for their medical needs. Efforts should be made to provide medical facilities in their respective areas so that they can receive timely attention and care. An integrated approach will help to improve health, life expectancy and quality of life of beta thalassemia patients.

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