Effectiveness Of Blood Transfusion Therapy And Awareness Among Kinfolks Of Thalassemia Patients Visiting A Tertiary Care Hospital In North India

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Introduction: The thalassemia is the most prevalent monogenic disease occurring at a high gene frequency throughout the thalassemia belt extending from Western Europe to Southeast Asia[1]. India, being ethnically diverse, frequency varies between several populations of different regions, averaging between 3-4%[2]. β-thalassemias are characterized by decreased synthesis of β globin chain of adult haemoglobin (HbA) resulting in transfusion dependant anaemia ,ineffective erythropoiesis, bone deformities with abnormal facies, hepatosplenomegaly and iron overload[3].

Although the only definitive cure for β-thalassemia is bone marrow transplantation, the mainstay of management still lies on blood transfusion therapy in Indian setting because of unaffordability of majority of the affected patients. The aim of present work is to study the effectiveness of transfusion therapy in thalassemia major patients as well as to assess the knowledge and awareness among the kins of the registered patients of our hospital.

The study was carried out in the Department of Transfusion Medicine, at a tertiary care hospital, New Delhi for a period of 6 months (February 2013 to July 2013) on 56 beta thalassemia patients (55= thalassemia major and 01 = thalassemia intermedia) visiting the hospital. A detailed history was taken through a thalassemia record proforma filled on recall basis by accompanying person or patient himself.

The history included demographic details, age at diagnosis, place of diagnosis, age at first transfusion, parents thalassemia screening status, history of transfusion from other hospital, type of blood component transfused, result of transfusion transmissible infections (TTI) screening, serum ferritin levels and whether leucofilter was ever used for transfusion.

Fifty six patients comprised of 35 males (62.5%) and 21 females (37.5%) in the age group of 07 months to 25 years (mean age = 11.06 years) were registered with the Thalassamia Day Care Centre but only 46 patients visited our hospital for blood transfusion during the study period.

Out of the total, 39 patients could provide necessary information as regards relevant past medical and transfusion records, diagnosis and immunization status records for inclusion into the study.

The blood samples for 46 patients were available for additional blood bank testing comprising red cell antibody screening, autocontrol testing and RH –Kell phenotyping. Recall by Mother: Father: Grandfather: Relatives was observed as 21:12:3:3. Ten (25.64%) patients were routinely coming from far off distances from neighbouring states after making suitable arrangements for travel and accommodation during each visit and three families had actually relocated to Delhi for blood transfusion purposes only. All of the patients had single earning member (father) in the family and the occupational profile was observed as small business/shopkeeper: private job: government job: driver: agriculturist: laborer to be 19:4:8:3:3:2 , highlighting the need for cost effective transfusion therapy for such patients.

Age at the time of diagnosis as confirmed by haemoglobin electrophoresis, high performance liquid chromatography or both for β-thalassemia ranged from 2 ½ months to 7 years (Table 1). All the patients were diagnosed at Dr. Ram Manohar Lohia Hospital or All India Institute of Medical Sciences, New Delhi except the 10 patients who were diagnosed at private hospitals in other cities.

Table 1: Age at the time of the diagnosis

Age	Number of patients
< 3months	07
3-6months	08
7-12months	09
1-2 years	08
3-7 years	07

The parents of 32 patients (82.05%) were reported to be screened for thalassemia trait. All the patients were

first transfused at the time of diagnosis except two who had received blood transfusion even before they were diagnosed as thalassemia major. Twelve patients gave history of blood (whole blood) transfusion from other private hospitals during their first episode of blood transfusion and later were registered with our hospital and were subsequently being transfused packed red cells only. Routinely, best efforts are made to provide inline filter leucoreduced red blood cells to all the thalassemia patients as per availability of the blood group. Pre-transfusion haemoglobin levels ranged from 4.9 g/dl to 8.8 g/dl (mean 6.69 ± 1.79 g/dl) .Those patients (10 in number) who were coming from far off areas often failed to maintain recommended haemoglobin levels due to their inability to abide by the set protocol for timely transfusion. Three such patients visiting once in 3 months for blood transfusion , presented to Thalassemia clinic with pre transfusion haemoglobin levels between 5-6g/dl.

During the study period ABO, Rh- Kell phenotyping, antibody screening and autocontrol testing were done on patients blood samples available at the blood bank as baseline investigations after obtaining informed consent.

ABO phenotype among registered thalassemia patients was observed to be B Rh D positive 24 (42.85%), O Rh D positive 14 (25%) , A Rh D positive 10 (17.85%), AB Rh D positive 05(8.92%) , B Rh D negative 02 (3.57%) and one A Rh D negative. Rh and Kell phenotyping was done on all the patients available during the study period before the scheduled blood transfusion and revealed 25 with DCcee, 14 DCcEe , 4 DCCee , 2 dccee and 01 dCcee phenotypes and 5 patients (10.87%) were found to be Kell antigen positive. None of the patients showed presence of any red cell alloantibody as tested by the three cell panel for antibody screening (Ortho Clinical Diagnostics Limited) unlike reported by other studies with small number of patient enrolment[4,5,6].

Three patients had positive autocontrol which did not interfere with compatibility testing and fully compatible crossmatched blood was issued to such patients. This gave us a preliminary record to plan for better match red cell transfusion therapy using appropriate antigen negative red cell unit from blood inventory to prevent alloimmunisation to the Rh (C ,c, E and e) and Kell antigens in future.

Only 19 parents / patients (48.17%) were aware of the TTI screening and three of them were updated about being reactive for one of the infectious marker. Due to ethical concerns, the TTI test results were not incorporated as part of the current study. Seven patients recalled use of bedside leucofilter for red blood transfusion in the past and others denied its use because of cost issues .The serum ferritin level ranged from 1100ng/ml to 6740 ng/ml (3943.37 ± 1931.31 ng/ml) in all the patients and the highest being observed in the oldest patient (25years) .

All the patients were on oral iron chelation therapy - Tab Desifer (taken as single dose per day) available as hospital supply. However, compliance to iron chelation therapy was needed to bring down toxic levels of serum ferritin in body. Number of blood transfusions per year per patient ranged from 4 – 36 as per the age and weight of the patients. Splenectomy was done in one patient in view of increased frequency of red blood cell transfusions and shortened interval between two transfusions.

Although the mild transfusion reactions were under-reported in our hospital setting, there was no history of acute hemolytic transfusion reaction being reported by the patients in the past. Rational use of leucoreduced saline adenine glucose mannitol(SAGM) suspended packed red blood cell has led to a decline in common acute transfusion reactions like febrile non hemolytic and allergic reactions. Moreover, absence of alloimmunization indicates the effectiveness of the blood transfusion therapy being practiced at our center. The socioeconomic status of the patient also play a vital role in quality care of the disease and a dedicated team of doctors, nurses and social workers is required for adequate management of thalassemia patient.

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