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Review Article

Provision of ideal transfusion support – The essence of thalassemia care

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ARTICLE INFO

Article history:
Received 21 July 2014
Accepted 28 July 2014
Available online 22 August 2014

Keywords:
Thalassemia
Transfusion support
Multitransfused

ABSTRACT

Thalassemia major is a major cause of transfusion dependence among patients world over. Provision of an adequate, uninterrupted and safe blood supply for these patients is the responsibility of the blood services as well as the society as a whole. Thalassemia management has evolved over a period of time and so have transfusion services. Various technological advancements have been introduced in the last few decades in order to enhance blood safety. Adoption of these newer technologies coupled with increasing awareness about voluntary blood donation in the general population can go a long way in improving the life expectancy as well the quality of life in these children.

1. Introduction

Thalassemia is an autosomal recessive disease prevalent in India. Even though efforts are being made at various levels to control this disease, the number of affected individuals remains huge. The therapeutic options available for management of thalassemia major are based on lifelong transfusion dependence and iron chelation.[1,2] Bone marrow transplants are now being promoted in advanced centres as a treatment modality, but the success rates in India are currently unknown.

Patients with thalassemia major are, therefore, largely transfusion dependant, forming the bulk of multiply transfused individuals in a population. It is the prime responsibility of a facility to offer its patients an adequate and uninterrupted supply of the safest possible blood for transfusion.

Thalassemia management has evolved over a period of time and so have transfusion services. Initially transfusions were given to these children only as a life saving measure, but the patients suffered due to poor quality of life and remained incapacitated. Orsini et al.[3] in 1961 and Wolman et al.[4] in 1964 demonstrated that "the results of Transfusion Therapy regularly and methodically repeated are absolutely superior to those achievable with transfusion given irregularly and only when the child appears quite anaemic".

2. Goals of transfusion

The aims of transfusion therapy are to correct the anaemia and maintain a circulating level of haemoglobin sufficient to suppress ineffective erythropoiesis, while minimizing iron

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http://dx.doi.org/10.1016/j.apme.2014.07.011

overload.[5] It is essential to maintain red cell viability and function during storage, to ensure sufficient transport of oxygen. At the same time avoidance of adverse reactions, including transmission of infectious agents is also important.

3. Embarking on transfusion therapy in thalassemics

Transfusion therapy should be started as soon as a diagnosis of thalassemia major has been established both on clinical and laboratory observations.[5]

There are certain pre-requisites before embarking on transfusion therapy. These include ABO & Rh (D) grouping of the patient along with extended phenotyping for minor blood group antigens like Kell, Kidd, Duffy, MNS antigens etc. Of these, the Rh (C, c, E, e) and Kell antigens are the most important since a large majority of antibodies reported in literature are directed against one of these antigens.[6] Thalassemics, by the virtue of being chronically transfused, are at a higher risk of developing alloantibodies.

4. Pre-transfusion testing

The usual transfusion policy is to perform ABO and Rh (D) typing of donors and patients and subsequent compatibility testing. However, there are many minor but clinically significant blood groups where alloimmunization may occur in multitransfused patients. Once the alloantibodies develop, finding compatible units may become difficult. Therefore, in addition to ABO and Rh(D) compatibility, blood matched for other minor antigens, especially from the Rh and Kell system should be preferred. Besides antibody screening should be repeated before each transfusion episode and if positive an attempt should be made to identify and characterize the antibodies. Thereafter it is important to always transfuse blood units lacking the antigens against which alloantibodies have developed.[6]

5. Basic requirements for transfusion

Transfusion medicine has evolved from a mostly laboratory – cantered service with a focus on serological aspects of blood, into a clinically oriented discipline that emphasises patient care. The implementation of safety and quality measures, progressively put forth during the last half-century, has subsequently improved the safety of blood and blood components. The aim of blood transfusion services should be to provide blood and blood products, which are as safe as possible, and adequate to meet patients' need and to maintain cut-off levels of blood and blood components in every Blood Bank to ensure blood availability in emergency.

Providing safe, adequate, timely and uninterrupted blood supplies to thalassemia patients is the responsibility of the blood centre under which the child is registered. But it is also the responsibility of the society as a whole to come forward and donate blood so that there is no shortage of this precious resource in the blood banks. Our society is becoming aware of this cause but unfortunately the gap between the demand and supply of blood in India is still very large.

Our transfusion services infrastructure is highly decentralized and lacks many critical resources; overall shortage of blood, especially from voluntary donors; limited and erratic testing facilities; an extremely limited blood component production/availability and use; and a shortage of health care professionals in the field of transfusion services.[7]

5.1. *Leucoreduced blood*

In spite of several advancements that have taken place, one thing has remained constant … ….."the requirement of the raw material- blood". The ideal product for transfusion in thalassemics is Packed Red Cells, preferably leucoreduced.

As per the Department of AIDS Control (DACS), Ministry of Health and family Welfare, Government of India, the proportion of blood components prepared was 41.3% in 2009–10 and the facility for component preparation is available only in select centres.[8] The concept of leucoreduction is further limited. Leucoreduction involves the removal of leucocytes from the blood components. The general norm is to use bedside leucodepletion filters as and when required. These do offer some protection but are not an ideal option for leucoreduction.

India has a population of one billion and has a huge burden of patient population requiring multiple transfusions. The population of chronically transfused individuals is increasing regularly with a rise in the number of hemato-oncological problems. Alloimmunization due to red cell, platelet or HLA antigens is a major problem associated with repeat transfusions. Therefore, transfusion of leucoreduced blood components assumes a lot of significance in these patients.[9]

Removal of leucocytes from various blood products has been shown to minimize Febrile non haemolytic transfusion reactions (FNHTR), HLA alloimmunization, platelet refractoriness in multitransfused patients and prevention of transmission of leucotropic viruses such as EBV and CMV. It also offers some protection against storage lesions, GVHD and immunomodulation.[10]

Keeping in view the variability of leukocyte numbers in the component and the leucoreduction method, the leukocyte content in a blood component unit should be less than 5×10^6/unit after leucoreduction (3 log reduction 99.9%) with a minimum of 85% red cell recovery in 95% of the units tested, as per the standards of the American Association of Blood Banks.[11] The European council guidelines are a little more stringent in terms of residual leukocyte content and require it to be less than 1×10^6/unit.[12]

Broadly leucoreduction is based on the principles of filtration of centrifugation. The various methods of leucoreduction include:

Pre-storage filtration of whole blood carried out with an in-line filter within 8 h after blood collection.

Pre-transfusion, laboratory filtration: Packed RBC prepared from donor whole blood then filtered prior to release from blood bank

Bedside filtration: Packed red cell unit is filtered at the bedside.