ABC of Blood Transfusion in Patients with Thalassemia Major

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ABSTRACT

Although blood transfusions are life sporting measures, it can cause alloimmunization, iron loading, bone marrow suppression, and infections etc. Therefore, it should be used according to patients' needs and blood groups should be dtermined prior to the first transfusion.

Key Words: Thalassemia, blood transfussion

INTRODUCTION

Among the hypochromic microcytic anemias, iron deficiency anemia, anemia of chronic inflammation, sideroblastic anemia,and thalassemia (homozygous or double heterozygous) become symptomatic in early infancy. Thalassemias are monogenetic hereditary disorders related to decrease in the synthesis of globin chains (alpha, beta, delta, and gamma). These are the most prevalent monogenic disorders in the world though mostly seen on the hemoglobinopathy belt. Patients with homozygous,double heterozygous condition present as microcytic hypochromic hemolytic anemia, but minimal hematologic findings might also be seen with heterozygous inheritance. In severe forms, hepatosplenomegaly, paleness, and jaundice usually develop in early life.

Among several types of thalassemias, alpha-thalassemias are the most prevalent form. In addition to alpha-thalassemias, beta-thalassemias, deltathalassemias, gamma-thalassemias, and their combinations are among the most common hereditary monogenic hematologic disorders in the world. Although alpha forms are the most prevalent thalassemias in the world (1), their extremely severe forms are seen mostly in intrauterine life or in a very early neonatal period with high mortality (2).

Beta-thalassemias, although not as frequent as alpha-thalassemias, generally are severe forms of diseases with extremely severe anemia and hepatosyplenomegaly developing in infancy. The severity of beta-thalassemias is mostly related to cis deletion of beta changes than the trans deletion. In addition to early hepatosplenomegaly and hypochromic severe anemia, normoblastemia is almost a pathognomonic form of the disease. Fetal hemoglobin concentration, especially the elevation of hemoglobin A2, is observed in individuals with beta-thalassemia trait and thus helps in the diagnosis of this disease. Severe thalassemias homozygous or double heterozygous are major forms in which patients survival, growth, and development depends on blood transfusions. Although this severe form is getting obsolete following prenatal diagnosis, it is still seen mostly in underdeveloped countries where prenatal diagnosis is less frequently performed. Thalassemia intermedia is less severe than thalassemia major;

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blood transfusion may be required for the intermedia form though less frequently than for the major form. Patients with thalassemia intermedia (3) and minima do not require transfusions regularly, though their findings are important for genetic evaluation.

Although blood transfusions (red cell transfusions) are life-saving in thalassemia major cases, they could also hamper the life of patients with the development of immune reactions, iron overloading, and transferring infections [hepatitis (A, B, C, and possibly E), HIV (especially in areas that are frequent), malaria cytomegalovirus, syphilis, etc.]. Therefore, before starting the first transfusion, these infections should be looked for in patients and in donors. In addition blood groups ABO, Rh (especially D, c, E, and C), Kell, Duffy, and other erythrocyte antigens should be determined as far as possible and be recorded on the first page of the patients chart. The donors blood should be selected according to the patients blood and subgroups as much as possible. Patient subgroups do not require to be determined after the initial examination unless transfusion reactions occur. ABO, D, c, E, Kell, Duffy antigens are the most important for immunization. Direct and indirect Coombs and ferritin determinations should be carried out before each transfusion and in the presence of iso- and auto-immunization.

Hematocrit (and also hemoglobin) should be determined in patients before each transfusion. Leukopheresis should be performed without irradiation. In each post-transfusion period, the patients hemoglobin level should be determined (at least 3 hours after the completion of transfusion). The hemoglobin level should be increased over 13 g/dL to suppress the patients bone marrow. If 13 g/dL is not reached, the rest of the same donor blood should be given the next day if it is left. When loading is suspected or observed, diuretic should be given to the patient and the transfusion should be discontinued immediately. When the post-hemoglobin level reaches over 13 g/dL, transfusion intervals should be longer and spleen should be checked to make sure it is reduced in size. Otherwise, the patient should be reevaluated before transfusion. This kind of high transfusion is important for growth in children, although it might increase iron overload. The iron overload can be overcome with iron chelation treatment, and administration of hepcidin or its agonist. For the prevention of febrile and lung symptoms, aspirin is recommended. During each transfusion, the reaction and its possible causes should be recorded on the patients chart. Intrauterine transfusions could be required in alpha- and gamma-thalassemias.

As blood transfusion can be dangerous although it is lifesaving, other approaches, such as stem cell transplantation, should be performed as early as possible. Some improvements may also be obtained by hypomethylation drugs. Of course, the best approach is the prevention of the birth of thalassemia major babies by premarital and prenatal diagnoses.

Although keeping hemoglobin level around 7 g/dl versus 10 g/dl does not affect patients prognosis with septic shock (10), keeping post transfusion hemoglobin level over 13 g/dl is important in thalasemic patients for their growth and decrease of splenic size.

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