Anemias Necessitating Transfusion Support

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Dear Editor,

The great benefit of blood/blood constitutes therapy is the ability to provide transfusion support for patients with many unique hematologic conditions. For some patients, such as patients with sickle cell disease, thalassemia major, immune hemolytic anemia, anemia of kidney disease, and aplastic anemia may need for this consolidation extends throughout their life. By knowing the alteration mechanisms of these conditions, we can appreciate the stationary, urgency, and the value of the transfused red blood cell (RBC).

Sickle cell disease is one of the prominent hemoglobinopathies. The anomaly is caused by the substitution of valine for glutamic acid at position 6 in the β-chain of the hemoglobin molecules. Hemoglobin S (Hb αβγδS) polymerizes under conditions of low oxygen tension, causing the sickling phenomenon. The clinical symptoms expressed by sickle cell patients indicate when RBC transfusion is necessary.

Sickle cell patients are highly exposed to potential risk for alloantibody production (20–30%), this may be due to chronicity of RBC support throughout most of his or her life. To avert this problem, partially phenotypes (D, E, C, c, e, and K) RBC of sickle cell patients should be matched.

Thalassemia is also a congenital disorder characterized by decrease synthesis or the formation of abnormal hemoglobin structure. Thalassemia syndrome is caused by a deficiency in α-chain or β-chain outputting. Complete lacking of α-chain is incompatible for life (fatal in utero), and absence of β-chain is considered as thalassemia major. Hepatosplenomegaly and bone marrow deformity may result due to hemopoietic tissue expansion to compensate for the anemia. RBC transfusion is indicated to suppress the ineffective hemopoiesis crisis and avoid the potential complications of the syndrome. The immune hemolytic anemia pertains to a group of disorders distinctive by short survival of erythrocyte due to an antibody (Ab) sensitization (coating) the red cell membrane. This sensitization triggers the red cell to be rapidly removed from the circulation much sooner than the normal red cells (life span, 120 ± 30 days). These disorders are identified as autoimmune hemolytic anemia (Ab reacts to self-antigen on red cell removed by spleen), drug-induced hemolytic anemia (drug and/or the Ab adsorb onto red cell), and alloimmune hemolytic anemia (RBC clearance for alloantibodies). In all forms, the identification of the causative agent should be considered the initial step in treatment. RBC transfusion is indicated, but depending on the extent of the anemia. Nevertheless, in patients with low hemoglobin but no clinical symptoms, the transfusion should be avoided.

Patients undergoing hemodialysis may have different hematologic complications that require transfusion therapy, often in the form of packed red cell support. Elevated level of blood urea leads to alterations in the red cell shapes, which prevent the red cell to traverse the spleen and removed prematurely by macrophage. In addition, the mechanism of dialysis itself causes a snipping of red cell, which also contributes to hemolysis. In this case, the recombinant erythropoietin use has significantly reduced the demand of transfusion; however, in severe anemia, RBC transfusion is indicated. Transfusion is usually started while the patient is on dialysis process to minimize the need for an additional venipuncture.

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Aplastic (hypoplastic) anemia or hemopoietic stem cell (HSC) disorder is defined as the simultaneous presence of pancytopenia, reticulocytopenia, and depletion of HSC resulting from aplastic bone marrow. It is categorized as primary (acquired or congenital) or secondary type. About 80% of the primary aplastic anemia is acquired, whereas 15–20% are congenital.\(^5\) Primary acquired aplastic anemia is further classified into idiopathic and secondary. Idiopathic acquired aplastic anemia is the most form (70%). Three inherited disorders have been included in the primary congenital form (Fanconi’s anemia, dyskeratosis congenital, and Shwachman-Diamond syndrome). The secondary aplastic anemia may result from direct damage to the HSC marrow by radiation or cytotoxic drugs. In case of aplastic anemia, blood product replacement should be provided to avoid alloimmunization, while the congenital form of aplastic anemia needs special circumstances.\(^6\) Pure red cell aplasia (PRCA) is a rare disease, distinct by a selective or severe decline in erythrocyte precursor simultaneously with normal marrow. The PRCA is classified into acquired or congenital. Acquired PRCA is categorized into acute or chronic form. Diamond-Blackfan anemia is the inherited erythroid hypoplastic form present in early infancy. RBC transfusion support is the mainstay in the PRCA, accompanied by some steroid drugs.\(^7\) Overall, bone marrow transplant improves outcome.\(^6\)

**REFERENCES**
