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Transfusion support in patients with sickle cell disease ☆

Deva Sharma [a,b], Ann Abiola Ogbenna [c], Adetola Kassim [a,e,*], Jennifer Andrews [b,d]

[a] Division of Hematology and Oncology, Department of Medicine, Vanderbilt University Medical Center, Nashville, TN, USA
[b] Division of Transfusion Medicine, Department of Pathology, Vanderbilt University Medical Center, Nashville, TN, USA
[c] Department of Hematology and Blood Transfusion, College of Medicine, University of Lagos, Lagos, Nigeria
[d] Division of Hematology and Oncology, Department of Pediatrics, Vanderbilt University Medical Center, Nashville, TN, USA
[e] Vanderbilt-Meharry Sickle Cell Center of Excellence, Vanderilt University Medical Center, Nashville, TN, USA

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ABSTRACT

Blood transfusions are an integral component of the management of acute and chronic complications of sickle cell disease. Red cells can be administered as a simple transfusion, part of a modified exchange procedure involving manual removal of autologous red cells and infusion of donor red cells, and part of an automated red cell exchange procedure using apheresis techniques. Individuals with sickle cell disease are at risk of multiple complications of blood transfusions, including transfusional hemosiderosis, auto- and alloimmunization to minor red cell and human leukocyte antigens, delayed hemolytic transfusion reactions, and hyper-hemolysis. In low- and middle-income countries in sub-Saharan Africa, where a directed donor system is prevalent and limited laboratory methods are in place to perform extended red cell phenotyping, leukodepletion of cellular products, and infectious disease screening, there are additional challenges to providing safe and adequate transfusion support for this patient population. We review current indications for acute and chronic transfusions in sickle cell disease that are derived primarily from randomized controlled trials and observational studies in children living in high-income countries. We will highlight populations with unique transfusion needs, such as pregnant women and children, as well as the role of the transfusion medicine consultative service for individuals with sickle cell disease planning to have curative hematopoietic stem cell transplantation or gene therapy. Finally, we will discuss risk factors for alloimmunization in individuals with sickle cell disease, emerging new strategies to prevent alloimmunization in this population, and critical gaps in the implementation of transfusion guidelines for sickle cell disease in high- and low-income countries.

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Introduction

Sickle cell disease (SCD) is an inherited blood disorder due to a point mutation in the beta globin gene, resulting in the production of insoluble sickle hemoglobin [1]. Numerous complications of vaso-occlusion can occur in affected individuals, including but not limited to recurrent acute pain episodes, silent cerebral infarcts and stroke, priapism, pigmentary cholelithiasis, functional asplenia secondary to repeated splenic infarcts and avascular necrosis [2]. Blood transfusion therapy is integral for the management of several acute and chronic complications of SCD [3]. Red blood cell (RBC) transfusions can be administered as a simple transfusion (infusion of donor RBCs), modified exchange transfusion that entails manual removal of autologous whole blood and infusion of donor RBC, and automated red cell exchange using apheresis methods (erythrocytapheresis). Although blood transfusion therapy can decrease SCD-related morbidity, it is associated with a number of risks, including alloimmunization, transfusional hemosiderosis, delayed hemolytic transfusion reactions, hyperhemolysis, and sensitization to donor human leukocyte antigens (HLAs) prior to curative hematopoietic stem cell transplantation [4]. In this review, we will discuss the current benefits, risks, and indications of blood transfusion therapy for the management of children and adults with SCD. We will also highlight opportunities for improvement in the administration of blood products for individuals with SCD, with a focus on emerging strategies to reduce the risk of alloimmunization.

☆ Co-last authors contributed equally to the writing of this manuscript.

* Corresponding author. Adetola Kassim, MD, Division of Hematology and Oncology, Vanderbilt University Medical Center, Nashville, TN 37232. Tel.: +1-615-936-8422; fax: +1-615-936-1812.
E-mail addresses: adetola.kassim@vumc.org, adetola.kassim@Vanderbilt.edu (A. Kassim).

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