Management of Patients with Sickle Cell Disease Using Transfusion Therapy

Guidelines and Complications

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KEYWORDS

• Sickle cell disease • Red blood cell transfusion • Alloimmunization • Iron overload

KEY POINTS

- Urgent or emergent red blood cell transfusion is indicated for acute ischemic stroke, acute chest syndrome, splenic or hepatic sequestration, transient aplastic crisis, multisystem organ failure, intrahepatic cholestasis, or obstetric complications in patients with sickle cell disease (SCD).
- Chronic transfusion therapy is indicated for primary and secondary stroke prevention and short-term for prevention of splenic sequestration recurrence.
- Patients with SCD should receive red cells antigen matched for C, E, and K to reduce alloimmunization risk.
- The iron status of chronically transfused patients with SCD should be closely monitored and iron chelation therapy and/or erythrocytapheresis implemented to maintain iron balance.

INTRODUCTION

Over the past few decades, significant advances in the care of patients with sickle cell disease (SCD) have led to improvements in morbidity and survival. The average life span of patients with SCD has increased from 14 years in 1973 to more than 50 years.[1] A key component in the management of patients with SCD is red blood cell (RBC)

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transfusion therapy. The major goals of RBC transfusions are relief of anemia, reduction of circulating sickle hemoglobin (HbS) erythrocytes, and improvement in oxygen-carrying capacity.[2] Although transfusion can be lifesaving, it is not without adverse effects. Using evidence-based transfusion policies can minimize transfusion-related complications. This review addresses RBC transfusion methods, indications (**Table 1**), and complications.

METHODS OF TRANSFUSION THERAPY

RBC transfusions can be administered by simple or exchange transfusion. Exchange transfusion is preferably performed by automated erythrocytapheresis but can be performed manually. Simple transfusions are dosed in units (1–3 units for adults) or

Table 1
Indications for transfusion therapy in adults and children with sickle cell disease

Transfusion Indication	Transfusion Method
Generally accepted indications for transfusion	
Acute ischemic stroke	Exchange transfusion preferred
Primary stroke prevention	Chronic simple or exchange transfusion[a]
Secondary stroke prevention	Chronic simple or exchange transfusion[a]
Acute chest syndrome (acute)	Simple or exchange transfusion[a]
Acute splenic sequestration	Simple transfusion
Acute splenic sequestration, recurrence	Chronic simple transfusion (before splenectomy)[b]
Preoperative (when general anesthesia required)	Simple transfusion
Transient aplastic crisis	Simple transfusion
Acute multisystem organ failure	Simple or exchange transfusion[c]
Acute hepatic sequestration	Simple or exchange transfusion[c]
Acute intrahepatic cholestasis	Simple or exchange transfusion[c]
Acute sickle or obstetric complications during pregnancy	Simple or exchange transfusion[c]
Controversial indications for transfusion	
Acute chest syndrome (recurrent)	Chronic simple or exchange transfusion[c]
Vasoocclusive painful episode (recurrent)	Chronic simple or exchange transfusion[c]
Pulmonary hypertension	Chronic simple or exchange transfusion[c]
Transfusion generally not indicated	
Uncomplicated vasoocclusive painful episode	NA
Priapism	NA
Uncomplicated pregnancy	NA
Leg ulcers	NA
Nonsurgically managed avascular necrosis	NA

Abbreviation: NA, not applicable.

[a] Exchange transfusion may be preferred in rapidly deteriorating patients when emergent HbS reduction is needed or when there are concerns for post-transfusion hyperviscosity due to a high pretransfusion hemoglobin (ie, >9 g/dL).

[b] Chronic transfusion may be used to delay but not prevent the need for splenectomy in very young children (ie, <2 years) who are at increased risk for invasive pneumococcal infections.

[c] Exchange transfusion may be preferred in patients with iron overload.