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Policies Specific to Patients with Sickle Cell Disease and Thalassemia-Blood Bank

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Document Type: Procedure

I. PURPOSE AND OBJECTIVE:

This document will provide policies and procedures that will enable the Blood Bank to provide red blood cells (RBCs) for patients with sickle cell disease (SCD) and thalassemia. Patients with sickle cell disease include those with sickle cell anemia, sickle cell β –thalassemia, and sickle cell hemoglobin C; refer to the *Scope* section for additional information. Patients with thalassemia include those with α -thalassemia or β -thalassemia.

II. INTRODUCTION:

Patients with sickle cell disease (SCD) and thalassemia are frequently transfused. The alloimmunization rate for chronically transfused patients with SCD is approximately 30%, compared with only 5% for other multiply transfused patients. Although the alloimmunization rate of thalassemia patients is not as high as SCD patients, they are also found to have a higher alloimmunization rate than other multiply transfused patients. The Blood Bank has adopted several policies to help address the needs of these patient populations and to help prevent alloimmunization.

III. SCOPE:

This document applies only to patients who the Blood Bank becomes aware of who have sickle cell disease or thalassemia including:

A. Sickle cell anemia: the homozygous state of HbSS.
B. Sickle cell β –thalassemia: the compound heterozygous state of HbS and β –thalassemia.

C. Sickle cell hemoglobin C:HbSC; the compound heterozygous state of HbS and hemoglobin C.

D. α -thalassemia or β -thalassemia, major or minor

This document **does not apply** to those patients possessing Sickle cell trait (inheritance of the heterozygous state of HbS).

If the technologist is uncertain whether the patient falls under the scope of this document, then a request for *Medical Review of Special Transfusion Requirements* should occur.

IV. POLICY:

A. Policy to Provide Hemoglobin S (HgbS) Negative Units to Patients with SCD and Thalassemia

RBC donor units for patients with SCD or thalassemia must be negative for hemoglobin S (HbS). Refer to, *Hemoglobin S Testing of Donor Units*.

B. Special Messages

If the Blood Bank becomes aware that a patient has SCD or thalassemia, then the HGBS "Use HbS negative RBCs" special message should be added to the computer record. If this message is added and the user attempts to issue a unit that is not hemoglobin S negative, then the user will receive a warning message: RBC units must be negative for the antigen(s). corresponding to any clinically significant antibody(ies).

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C. Policy to Provide Fresh RBC Units for Transfusion

All RBC units intended for transfusion (including exchange transfusion) to patients with SCD or thalassemia should be the freshest units available. If the anticoagulant is

1. CPDA, then the unit should not expire within 21 days (preferred) or within 14 days (if necessary).
2. Adsol, then the unit should not expire within 28 days (preferred) or within 21 days (if necessary).

If a banded pre-transfusion sample for a sickle cell disease or thalassemia patient consists of a T&S only, then two RBC units will be crossmatched per policy, regardless of whether the patient has alloantibodies.

D. Obtaining the Patient History

If the Blood Bank becomes aware that a patient has SCD or thalassemia then a patient history should be obtained. The reasons for obtaining this history include the following:

1. to help determine whether the patient is historically alloimmunized, as there is the potential that the antibody levels may drop below detectable levels.

2. the history may also reveal whether the patient has been transfused in the last 90 days.
3. contact with American Red Cross (ARC) and Versiti Reference Laboratories should occur to determine if either of these laboratories has any previous history or complete phenotype on file for the patient.

E. Policy to Provide Partially Phenotypically Matched RBCs for Transfusion for the C, E, and Kell Antigens

1. RBCs that are transfused to patients with SCD or thalassemia shall be preemptively matched to the patient/recipient for the C, E, and Kell antigens in an attempt to prevent alloimmunization to these highly antigenic RBC antigens.
2. When the Blood Bank becomes aware of a new SCD or thalassemia patient, sickle cell issue codes will be added to the patient's antibodies in SOFT to ensure proper antigen negative units are provided. The sickle cell issue codes are SIC_C, SIC_E, and SIC_K. Patients will have one, two, or all three codes added based on their antibody and antigen history.
    a. **SIC_C** is added to all SCD and thalassemia patients initially (regardless of antigen type) unless a true anti-C antibody has been formed. If the patient is positive for the C antigen, and the genotype report states there is no C variant, the SIC_C may be removed. See the *Genotype Testing* section below.
    b. **SIC_E** is added to all SCD and thalassemia patients that are E antigen negative or unknown, unless a true anti-E has been formed.
    c. **SIC_K** is added to all SCD and thalassemia patients that are Kell antigen negative or unknown, unless a true anti-Kell has been formed.
3. If the Blood Bank is unable to type the patient's RBCs, then donor units must be negative for the C, E, and K antigen(s).
4. When antigen typing patient samples or donor units, the technologist must also comply with the policies and procedures for antigen typing.

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F. Policy for Babies of SCD or thalassemia Moms

1. Babies of SCD/thalassemia Moms will have SIC codes matching the mother's SIC codes added to their record in SOFT.
2. An antigen negative Baby Unit, corresponding to added SIC codes needs to be ready and available.
3. SCD babies will be E and Kell antigen typed as sample allows. We will not request an additional sample for antigen typing.

G. Genotype Testing

1. All SCD and thalassemia patients will have a sample sent to Versiti Wisconsin for molecular genotyping, unless a molecular genotype is already on file for that patient.
    a. Patient will be billed for this test once the final results are entered into SOFT.

2. Upon receiving the molecular report back from the reference lab, the SIC codes will be removed from the patient record if the patient is positive for the corresponding antigen, and there is no variant for that antigen. If there are Rh variants in the report, or the technologist is unsure which SIC codes to remove, the molecular report will be brought to rounds for clarification.
3. **Examples of C, E, K Matching**
    a. *The Blood Bank antigen types a SCD patient who was last transfused five months ago; the patient's RBCs type C+ E+ K-. The Blood Bank shall provide RBC units that are C- and K- until molecular genotyping is performed to rule out the presence of C antigen variant. This patient will receive C- K- HbS negative RBCs. Once the molecular genotype is completed on this patient, the SIC_C code will be removed if there are no C antigen variants present. From that point on, the patient will no longer need C antigen negative RBCs.*
    b. *A new SCD patient requires a RBC transfusion. The Blood Bank obtains the patient history and learns that the patient has sickle cell β –thalassemia, was transfused two weeks ago, and also has a history of Anti-Fy$^{a.}$ The Blood Bank shall provide RBC units that are C- E- K- and Fy^{a}-. The units should also be fresh and HbS negative.*
    c. *A new SCD patient requires an immediate RBC transfusion. The patient history reveals that the patient was transfused nine days ago at another hospital. Because the Blood Bank cannot antigen type the patient (due to the recent transfusion), C- E- and K- RBCs shall be provided. The units should also be fresh and HbS negative.*

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V. NOTES:

A. If a SCD or thalassemia patient is C negative and therefore requires C negative RBC units, it may be helpful to use Rh(D) negative units. The incidence of the C antigen is lower in the Rh(D) negative population than in the Rh(D) positive population.
B. Sickle cell disease or thalassemia alone is not an indication for irradiation. If the technologist is uncertain whether a SCD/thalassemia patient requires irradiation components for any other reason, a *Medical Review of Special Transfusion Requirements* should occur.

VI. REFERENCES:

1. American Association of Blood Banks, *Technical Manual*, current edition.
2. *Blood Banking and Transfusion Medicine Basic Principles & Practice*, Hillyer, Silberstein, Ness, Anderson, and Roback, second edition, 2007.
3. College of American Pathologists Transfusion Medicine, TRM.40720, *Immunohematologic Conditions*, 2010.
4. Datta, S. S., Mukherjee, S., Talukder, B., Bhattacharya, P., & Mukherjee, K. (2015). Frequency of Red Cell Alloimmunization and Autoimmunization in Thalassemia Patients: A Report from Eastern India. *Advances in Hematology*, *2015*, 610931. http://doi.org/10.1155/2015/610931
5. Davoudi-Kiakalayeh, A., Mohammadi, R., Pourfathollah, A. A., Siery, Z., & Davoudi-Kiakalayeh, S. (2017). Alloimmunization in Thalassemia Patients: New Insight for Healthcare. *International Journal of Preventive Medicine*, *8*, 101. http://doi.org/10.4103/ijpvm.IJPVM_246_16

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