Transfusion Medicine Quality Manual
Guidelines for the Selection of Red Blood Cells for Transfusion of Patients with Sickle Cell Disease

1.0 Policy Statements

1.1 Physician/Designate shall, whenever possible, notify the transfusion medicine laboratory when there is a diagnosis of sickle cell disease.

1.2 Patients with sickle cell disease (SCD) shall have extended blood group phenotypes for ABO, D, C ,c, E, e, K, Jka, Jkb, Fya, Fyb, S and s performed prior to initial transfusion if possible.

1.3 Red cells selected for transfusion shall be phenotypically matched for at least the D, C, c, E, e and Kell antigens.

1.4 Red cells selected for transfusion to a recipient with sickle cell disease who has been identified with, or has a history of, alloantibodies shall be as phenotypically similar as possible for all the major blood group antigens.

1.5 Recipients with a known alloantibody shall receive red cells which lack the corresponding antigen.

1.6 Red cells selected for transfusion shall be tested negative for hemoglobin S whenever possible. (Sickledex).

1.7 Sickle trait blood should not be transfused to a patient with SCD.

1.8 Use of a cell saver is contraindicated in patients with SCD.

1.9 Autologous blood is contraindicated in patients with SCD.

1.10 Patients sample should be sent to the National Immunohematology Reference Laboratory for genotyping if phenotypes have not been performed and:

1.10.1 the patient has been transfused in the last three (3) months; and/or

1.10.2 antigen typing by the indirect antiglobulin test (IAT) are invalid if the patient has a positive direct antiglobulin test (DAT) with IgG. IgG coating on the red cell can sometimes be removed by treating the cells with glycine/EDTA (i.e. EGA) or chloroquine. If the DAT is negative on the treated cells they can then be used for phenotypings.

1.11 Special transfusion requirements shall be retained permanently in the patients' blood bank history.

1.12 Physician/Designate shall be consulted if the red blood cells for transfusion:

1.12.1 are **not** matched for at least D, C, c, E, e, and Kell antigens; or

1.12.2 are not screened for Hemoglobin S.

Note: Follow facility policy for emergency release of red blood cells if testing is not complete and patient requires immediate transfusion.

2.0 Linkages

Guidelines for Transfusion Orders for Blood Components and Blood Products: Available at:
http://www.health.gov.nl.ca/health/bloodservices/pdf/transfusion_orders_version_2.pdf

Merriam-Webster dictionary. Available at:
http://www.merriam-webster.com/dictionary/sickle-cell%20trait

Product information for Sickledex: available at:
http://www.streck.com/resources%5CHematology%5CSICKLEDEX%5C02_Product_Information%5C02_Paper_SICKLEDEX_-_Screening_Test_Guidelines.pdf

Standard Operating Procedure for ABO Grouping Tube Method. Available at:
http://www.health.gov.nl.ca/health/bloodservices/pdf/sop_for_abo_grouping.pdf

Standard Operating Procedure for Antigen Typing. Available at;
http://www.health.gov.nl.ca/health/bloodservices/pdf/antigen_typing_final_ver_1.pdf

Standard Operating Procedure for Patient History Check: Available at:
http://www.health.gov.nl.ca/health/bloodservices/pdf/patient_history_check.pdf

Standard Operating Procedure for Performing the Direct AntiglobulinTest**.** Available at:
http://www.health.gov.nl.ca/health/bloodservices/pdf/performing_the_direct_antiglobulin_test_ver3.pdf

Standard Operating Procedure for Quality Control of Reagents and Antisera
http://www.health.gov.nl.ca/health/bloodservices/pdf/quality_control_of_reagents_and_antisera_ver1.pdf

Standard Operating Procedure for Rh Typing Tube Method. Available at:
http://www.health.gov.nl.ca/health/bloodservices/pdf/sop_for_rh_typing.pdf

3.0 Scope

3.1 All Transfusion Medicine Laboratory Technologists.

3.2 All health care professionals who provide care and treatment to patients with Sickle Cell Disease.

4.0 General Information

4.1 Patients with SCD have the highest rate of red cell alloimmunization of any patient group.

4.2 Patients with SCD have elevated blood viscosity which may be exacerbated by increases in hematocrit.

4.3 Patients with SCD are more likely to experience delayed hemolytic transfusion reactions.

4.4 In the absence of symptoms, transfusion should be avoided until the hemoglobin is < 50 g/L.

4.5 In chronically transfused children with SCD, the goal of transfusion should be to maintain a HbS level of below 30% immediately prior to the nest transfusion.

4.6 Patients with SCD should not be transfused if the hemoglobin is > 100 g/L.

4.7 Matching for Fyb in SCD patients who are Fya negative and Fyb negative is not normally necessary unless the patient has an allo anti-Fyb.

4.8 In emergency situations blood may be transfused to SCD patients prior to sickle cell testing (Sickledex).

4.9 Fresh blood cells (< 14 days old) may deliver oxygen more efficiently.

4.10 CMV negative or irradiated products are not necessary unless the patient has a co-existing medical condition with these special requirements.

4.11 Iron overload complications are expected to occur after the transfusion of twenty (20) to thirty (30) units of red blood cells.

5.0 Process

5.1 Quality Control

5.1.1 Perform quality control on all antisera used with a positive and negative control. The positive control shall be heterozygous for the antigen in question whenever possible. If a heterozygous positive control is not available, any cell positive for the antigen can be used.

5.1.2 Record lot number, panel cell number and expiry date of all cells chosen for positive and negative controls.

5.1.3 Reagents shall be used according to manufacturer's instructions.

5.1.4 The negative control must lack the antigen in question.

5.1.5 Positive and negative controls must be performed each day of use.

5.1.6 If more than one lot number of antisera is being used for the same antigen, each lot number must have a set of positive and negative controls.

5.1.7 IgG coated cells must be added to all negative indirect antiglobulin test results to confirm AHG is present and working.

5.1.8 Sickledex kit is to be controlled according to manufacturer's instructions.

5.2 Procedure

5.2.1 Check patient's history for transfusions and previous phenotyping results.

5.2.2 Phenotype patients red cells for E, e, C, c, K, Jka, Jkb, Fya, Fyb, S, and s if no phenotypes have been done AND the patient has not been transfused in the last three (3) months,

Note: If the patient has a positive DAT due to IgG, antigen typings performed by IAT are invalid.

5.2.3 Crossmatch red blood cells (RBC's) that are phenotypically similar for:

5.2.3.1 D, C ,c, E, e and Kell antigens if the patient does not have a clinically significant antibody or a history of a clinically significant antibody; or

5.2.3.2 D, C, c, E, e, K, Jka, Jkb, Fya, Fyb, S,and s antigens if the patient does have a clinically significant antibody or a history of a clinically significant antibody.

5.2.4 Screen the donor unit for hemoglobin S (if possible).

5.2.5 Consult with physician/designate if recommended RBC's are not available.

5.2.6 Issue RBC's as per facility policy.

5.3 Guidelines

Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014: Retrieved from: **http://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines**

5.4 Materials

5.4.1 **Reagents:**

5.4.1.1 Polyspecific AHG containing anti IgG, C3d

5.4.1.2 Antisera (specific to the antigen in question)

5.4.1.3 Coombs control cells

5.4.1.4 Positive and negative control cells

5.4.1.5 Patient or donor red cell suspension

5.4.1.6 Sickledex reagent and appropriate controls

5.4.2 **Supplies:**

5.4.2.1 Transfer Pipettes

5.4.2.2 Test tubes (10 x75mm or 12x75mm)

5.4.2.3 Timer

5.4.2.4 Manufacturer's instructions specific to the antisera being used

5.4.2.5 Isotonic saline

5.4.3 **Equipment:**

5.4.3.1 Serological centrifuge

5.4.3.2 Refrigerator with temperature between 1-10◦C

5.4.3.3 Cell washer (if used at your facility

5.4.3.4 37(±1) °C waterbath or incubator

6.0 Acronyms

SCD	Sickle cell disease
CMV	Cytomegalovirus
RBC's	Red blood cells
AHG	Antihuman globulin

7.0 Definitions

Sickle cell disease	Sickle cell disease (SCD) is caused by inherited mutations involving the beta globin gene that result in the formation of an abnormal hemoglobin (hemoglobin S). Red blood cells, which contain a predominance of hemoglobin S, undergo shape change when low oxygen concentrations cause polymerization of the sickle hemoglobin. The damaged red blood cells become rigid and inflexible, occluding blood vessels and inducing tissue ischemia, pain, and organ damage. This process is accompanied by an inflammatory response and shortened red blood cell survival. These alterations may result in a wide variety of clinical manifestations
Sickle cell trait	a usually asymptomatic blood condition in which some red blood cells tend to sickle but usually not enough to produce anemia and which occurs in individuals (as those of African or Mediterranean descent) who are heterozygous for the gene controlling hemoglobin S.

hemoglobin S	Abnormal hemoglobin that occurs in the red blood cells in sickle-cell anemia and sickle-cell trait.

8.0 Records Management

8.1 The recipient transfusion data file in the transfusion medicine laboratory shall be retained indefinitely.

8.2 All transfusion records in the recipient's medical chart, including pre-transfusion serological tests results and worksheets for identification of atypical antibodies shall be retained in accordance with health care facility's retention policy for medical records.

8.3 Quality control of blood components, blood products, reagents and equipment shall be retained for 5 years.

8.4 Date and time of specimen collection and phlebotomist's identification shall be retained for 1 year.

8.5 Request form for serologic tests shall be retained for one month or as per facility policy.

8.6 Documentation of staff training and competency must be kept for a minimum of ten years.

8.7 Temperature monitoring records for blood products must be kept a minimum of five years.

8.8 Records of blood components inspection prior to release must be kept for a minimum of five years.

9.0 Key Words

Sickle cell disease

10.0 Supporting Documents

10.1 Process Flow/Algorithm (NA)

10.2 Tables/Charts (NA)

References

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