Government of Newfoundland and Labrador

Department of Health and Community Services
Provincial Blood Coordinating Program

SELECTION OF RED BLOOD CELLS FOR PATIENTS WITH SICKLE CELL DISEASE	NLBCP-056

Office of Administrative Responsibility	Issuing Authority
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Overview

Red blood cell (RBC) transfusions are a crucial treatment for patients with Sickle Cell Disease (SCD) and as a result these patients have the highest rate of RBC alloimmunization of any patient group. These patients sometimes have complex and partial antigen expressions on their red blood cells (RBCs) and therefore are likely to experience delayed hemolytic transfusion reactions.

It is important for the Transfusion Medicine Laboratory (TML) to be notified as soon as possible when a patient has SCD and may require a transfusion. The TML will obtain an extended RBC antigen profile at the earliest opportunity so that phenotypically matched RBCs can be provided if required.

Policy

1. The most responsible physician shall whenever possible notify the TML when there is a diagnosis of SCD.
2. The diagnosis of SCD shall be entered into the patient's TML permanent history.
3. SCD patients shall have phenotyping for antigens D, C, c, E, e and Kell performed, if possible, to determine the patient's phenotype before RBC cell transfusion. Antigens Fya/Fyb, Jka/Jkb, S/s should also be determined if possible.
4. A blood sample shall be sent to the National Immunohematology Reference Laboratory for a full extended RBC antigen profile so any partial variants/limited expressions can be identified.
5. RBCs selected for transfusion shall be phenotypically matched for at least antigens D, C, c, E, e and Kell if possible.
6. RBCs selected for transfusion to a recipient with SCD who has been identified with or has a history of alloantibodies shall receive RBCs that lack the corresponding antigen and should also be phenotypically matched for all other major RBC antigens.
7. If phenotypically antigen matched RBCs are not available, the physician should be notified.
8. RBCs for SCD patients shall be screened for Hemoglobin S if possible.

   Note: RBC units that test positive for Hemoglobin S are from donors with sickle cell trait. These RBCs would be safe for transfuse to SCD patients, however, would cause incorrect post-transfusion Hemoglobin S percentage. Transfusion of these RBC units should be avoided.

9. The prescriber shall be notified if the patient must be issued a Hemoglobin S positive RBC unit.

Guidelines

1. Patients who have been transfused RBCs in the last three months are not suitable for phenotyping.
2. A large majority of SCD patients that phenotype as Fyb-negative are actually Fyb-positive by genotype because of limited expression of antigens on their RBCs. These patients can be safely transfused Fyb positive RBCs.
3. The most responsible prescriber should be supplied a copy of the RBC genotype report so that the report can become part of the patient's permanent record.
4. Nearly one quarter of SCD patients will express only a partial form of the C antigen and therefore may be at risk of forming an anti-C antibody.
5. Patients with SCD that are not prophylactically transfused phenotyped RBCs can have alloimmunization rates as high as 19 to 43 percent with about 30 to 50 percent of patients having an existing antibody being undetectable within a year.
6. Exchange transfusions are required for some SCD patients.

Key Words

Sickle Cell Disease, Hemoglobin S, phenotype

References

Callum, J. L. (2022). *Bloody Easy 5.1: Blood transfusions, blood alternatives and blood transfusion reactions*. Toronto: Ontario Regional Blood Coordinating Network (ORBCoN).

Cohn, Claudia S (2023). AABB. *Technical manual*. 21st Edition. Maryland, USA: Author