

Red blood cell transfusion in sickle cell disease

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1 Red blood cell (RBC) transfusions are a life-saving therapy for patients with sickle cell disease

Most patients with sickle cell disease will require RBC transfusions. At a Canadian centre, 60% of patients received 1 or more RBC transfusions per year, with a subset receiving more than 100 RBC units per year.¹ Transfusion can improve oxygen delivery and decrease clinical features of vaso-occlusion.²

2 Reduction in the proportion of hemoglobin S, rather than increasing total hemoglobin, is often the target of transfusion

With physiological compensation for anemia in patients with sickle cell disease, transfusion based on hemoglobin threshold alone is avoided unless severe anemia (hemoglobin \leq 50 g/L) is present.² Transfusion increases blood viscosity, impairing oxygen delivery when hemoglobin exceeds 100 g/L.³ Exchange of RBCs may be required to avoid exceeding this level with simple transfusion.

3 Extended RBC antigen profiling improves safety of transfusion

Two-thirds of patients receiving transfusions regularly have evidence of RBC alloimmunization.⁴ Extended RBC antigen profiling should be performed before the first transfusion. Transfused RBCs should be matched for Rh (D, C, c, E, e) and Kell antigens.²

4 Transfusion and allo-antibody history should be obtained

Titres of RBC antibody can decrease over time to levels not detected in a type and screen test.⁵ With re-exposure to RBC antigen, delayed hemolytic reactions, including hyperhemolysis with life-threatening anemia can occur.⁵ A comprehensive transfusion history, including location and antibody history, will assist the blood bank in selecting appropriate RBCs for transfusion. Patients may carry a card with their RBC phenotype and allo-antibody history.

5 Consult with transfusion medicine and hematology before starting transfusion for each patient with sickle cell disease

Consultation ensures that indications and method of transfusion (simple v. exchange transfusion) are appropriate. Advance notice allows time for testing before the transfusion to provide the most appropriate products. The RBCs of the required phenotype may not be in routine inventory. However, in life-threatening circumstances, transfusion should not be delayed for RBCs that are matched by phenotype.

References

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