Sickle Cell Disease: Chronic Blood Transfusions

There may be times when sickle cell patients require a blood transfusion. Such situations include preparing for surgery, during pregnancy, or during a severe complication such as an aplastic crisis, splenic sequestration or acute chest syndrome. In these cases, transfusion is a one-time intervention used to reduce the severity of the complication you are experiencing.

However, if you have had a stroke, or an MRI or TCD shows that you are at high risk for having a stroke, your hematologist may recommend you begin **chronic blood transfusions.**

What Does a Blood Transfusion Do?

Chronic (monthly) blood transfusions have been proven to drastically reduce a sickle cell patient's risk of stroke. They have also been shown to reduce the frequency, severity and duration of other sickle cell complications. Sickle cell patients usually have a hemoglobin S level of about 80-90%. This means 80-90% of the circulating red blood cells are cells that can sickle and cause complications. The goal of chronic blood transfusion therapy is to bring that percentage down below 30%. This will mean fewer sickle cells circulating in the body, and a lower risk of complications from sickling.

How is Blood Given?

Blood can be transfused in two ways: a simple transfusion or a red blood cell exchange transfusion.

A *simple transfusion* involves transfusing 1-3 units of blood through a peripheral IV. The transfusion takes about 3-4 hours, and must be given every 4 weeks to help bring down the hemoglobin S to target levels. Unfortunately these blood transfusions can cause iron overload. After a year, the patient must begin iron chelation therapy to prevent organ damage *(see pamphlet "Iron Chelation")*.

An *exchange transfusion* is when a special machine is used to remove the patient's blood through an IV while simultaneously infusing healthy donor blood through a second IV. A red cell exchange takes about 3-4 hours and can bring hemoglobin S levels down to less than 10%. In addition, red cell exchanges do not cause iron overload, so no chelation is required. However, blood work must be done 2-3 days in advance, and IV access can be very difficult because of the size of IV required for the procedure.

Both types of transfusions have been proven to be effective at reducing the risk of stroke in sickle cell patients.

What are The Risks?

Blood transfusions are not without risks. One risk is *alloimmunization*, a process in which the patient receiving blood transfusions creates antibodies to certain types of blood. As a result he/she may have a reaction to the blood that was transfused. Alloimmunization makes it more difficult to find blood that is a good match for the patient. In order to prevent alloimmunization, some centers routinely perform RBC phenotyping (special testing for antibodies) on sickle cell disease patients so that they may give blood that is a better match for the patient. However, this extended testing still does not cover all antibodies, and reactions may still occur.

Another risk of transfusion is *hyperhemolysis*. Hyperhemolysis occurs when a patient's body destroys both newly transfused cells and the patient's own cells. In this situation, the hemoglobin level falls dramatically and the patient becomes very anemic. Hyperhemolysis is rare, but must be treated immediately to avoid becoming life threatening.

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