Chronic Transfusions

Chronic transfusion therapy occurs when a patient receives a blood transfusion once a month for many years. Chronic transfusions allow normal red blood cells (RBCs) to live longer in the body and flow freely in blood vessels.

The most common reason why children with sickle cell disease (SCD) are placed on chronic transfusion therapy is to prevent stroke (or repeat stroke) from occurring. Chronic transfusions may also be used for a period of six months to two years to prevent further episodes of other severe sickle-related complications (i.e. acute chest syndrome, splenic sequestration).

Children with SCD have sickle hemoglobin S (HbS) in their red blood cells. Chronic transfusions decrease sickle cell related complications by decreasing the amount of HbS. This is done by giving a blood transfusion to maintain the HbS at 30 to 50 percent. In order to maintain the HbS level within this range, transfusions are usually repeated every three to four weeks. Children with SCD who have had a stroke or are determined to be at high risk of having a stroke by Transcranial Doppler screening tests often receive chronic transfusions for many years.

Other Options

Hydroxyurea and stem cell (bone marrow) transplants are additional treatment options. Hydroxyurea has been proven to control many symptoms of sickle cell disease. Stem cell transplants can cure sickle cell disease, but require a suitable donor. Severe side effects can occur.

For information about the Transfusion Buddy Program at Children’s National, call (202)476-KIDS (5437)

For questions regarding the chronic transfusion program, contact: Jennifer Webb, M.D. jwebb@ChildrensNational.org
Primary Stroke Prevention

Sickle cell disease (SCD) is the most common cause of childhood stroke. The majority of strokes in this population occur between the ages of three and 15 years old. The annual risk of stroke in children with SCD is 0.6 to one percent per year. This rate is approximately 300 times higher than in children without SCD. Transcranial Doppler (TCD) screening should be done every year in children with SCD starting at age two. If the results of the TCD are abnormal, your child has a risk of stroke as high as 40 percent within three years of the abnormal TCD without treatment. Chronic transfusions given every three to four weeks reduce the stroke risk in children with SCD and abnormal TCDs by more than 90 percent.

Prevention of Recurrent Stroke

Children with SCD who have had one stroke are at extremely high risk of having another stroke. Research has shown that chronic transfusion therapy reduces the risk of additional strokes. Without chronic transfusion therapy, approximately 67 percent of children with SCD who have had a stroke will suffer from another stroke. However, with chronic transfusion therapy, the risk of recurrent stroke rate falls to less than 20 percent. The highest risk of having another stroke is in the first five years. Your child’s hematologist will determine what the target level of HbS they want to keep your child at, which is usually 30 percent.

Types of Transfusions

Simple Transfusion. Blood is given to the patient through an IV (most common).
Partial Manual Exchange Transfusion. Blood is first removed through an IV, then replaced with a blood and saline combination to ensure safe blood counts and volume (for high hemoglobin levels).
Automated Exchange Transfusion - Blood is removed and replaced with blood from healthy donors (done through IVs or special central lines) using an apheresis machine.

Risks of Transfusions

Iron overload
Excess iron from transfused red blood cells can build up and remain in the body. If left untreated, this iron can harm the liver, heart, and other organs. Your child’s hematologist will monitor for iron overload through monthly lab tests and yearly MRI scans of the liver. After about one to two years of chronic transfusions, patients often need to be treated with medicines to help remove iron from the body (iron chelators).

Infectious disease transmission
Every effort is made to ensure that the blood your child receives is safe of infectious agents. Donors are interviewed to make sure they are healthy, and every unit of blood is tested for known transfusion-transmissible infections. All blood is screened for viruses and other infections, including those that cause hepatitis, human immunodeficiency virus (HIV), human T-cell lymphotropic virus (HTLV), and syphilis. As a result, the chance of becoming infected with one of these viruses is extremely small.

Risks of Transfusion, Continued

Alloimmunization:

When a child receives blood that has different proteins (antigens) from their own, they may develop antibodies against these foreign antigens. This condition is called alloimmunization. People who are transfused frequently, such as patients with sickle cell disease, can form one or more antibodies against RBCs. This can make it very difficult to find blood for the patient. Rarely, reactions can occur in patients with antibodies bringing on fevers, muscle aches, joint pain, anemia, or dark urine.

At Children’s National the focus is on preventing the development of antibodies in several ways. Prior to each transfusion (within three days) the blood bank screens your child’s blood for any new antibodies that the child may have developed. We try to decrease the amount of antibodies SCD patients develop by closely matching the blood donor type to the patient’s type. The Blood Donor Center at Children’s National has established the Transfusion Buddy Program to provide our chronically transfused SCD patients with antigen matched blood products, which dramatically decreases the rate of alloimmunization.

Despite the risks, transfusions can provide substantial benefits to children with SCD.