Glossary

Anemia

Refers to having low hemoglobin or not enough red blood cells. There are many causes of anemia, including sickle cell disease, iron deficiency, bleeding, and many other genetic or acquired causes.

Fetal Hemoglobin (HbF)

The type of hemoglobin produced by babies before they are born. This hemoglobin is special and allows the baby to use oxygen from the mother's circulation. After the baby is born, this is no longer needed and the levels of HbF go down usually in the first 6-12 months of life. HbF protects against the complications of sickle cell disease, which is why young infants have no sickle cell disease complications early in life. HbF levels are closely monitored on hydroxyurea therapy with a goal HbF of at least 20-30%. The higher the HbF value, the more protected you are against the complications of sickle cell disease.

Hemoglobin

Hemoglobin is an important blood count for people with sickle cell disease. Without treatment, the normal hemoglobin for people with HbSS or HbS-Beta-Zero Thalassemia is ~7-8 g/dL. With hydroxyurea, we hope to increase the hemoglobin to levels greater than 10 g/dL. Patients with HbSC or other less severe forms of sickle cell disease typically have hemoglobin levels 10-12 g/dL. People without sickle cell disease have hemoglobin levels of 12-16 g/dL, depending on age and gender.

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Nuetrophils

The type of white blood cells that specifically fight bacterial infections. The absolute neutrophil count (ANC) is closely followed with hydroxyurea therapy. The goal is to reduce ANC to a safe range where the effects of hydroxyurea are maximized without any increased risk of infection. Routine blood draws (CBCs) are done to closely monitor the ANC.

Red Blood Cells

Red blood cells are among the most abundant cells in your body. They are responsible for carrying oxygen through your body in the blood stream. The normal lifespan of red blood cells is ~100-120 days. For people with untreated sickle cell disease, the lifespan of red blood cells is ~20 days. This is one of the key reasons why people with sickle cell disease have anemia.

Reticulocyte Count

Refers to the number of young red blood cells. When red blood cells are made in your bone marrow, they initially enter your circulation as reticulocytes. Because red blood cells do not live as long in patients with sickle cell disease, there are many reticulocytes being produced to try to “keep up” and prevent worsening anemia. With hydroxyurea treatment, the red blood cells are healthier and live longer so the reticulocyte count goes down close to normal.

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White Blood Cells

The type of blood cells that fight infection. For patients with sickle cell disease, the white blood cells are often elevated even if there is not an infection. Hydroxyurea will usually reduce the white blood cell count. Routine blood draws check to be sure that the white blood count is not too low.