**Literature Review:**

How has treatment for other blood diseases shown hope as possible treatment for sickle cell?

Sickle cell anemia, when left untreated, can become a lethal to the human body. The many irregular functions this disease causes leads to other diseases and/or complications such as pulmonary hypertension, stroke, and acute chest syndrome.

Many sickle cell patients unfortunately fall victims to stroke due to the sickle cells clogging the arteries that lead to the brain (What is Stroke?). Because sickle cell in known for the sickle cells blocking arteries, strokes are common among patients; specifically in children. If the patient is not brought to medical attention in a considerable amount of time, there could be significant brain damage in development, and/or daily activities such as walking, talking, or even lead to death. Blood Transfusions are common treatment for strokes, but as seen in Cheryl, blood transfusions can often lead to more harm than good. Blood transfusions are commonly used for sickle cell patients including spleen enlargement and anemia causing the low blood count. Constant blood transfusions cause the veins to become weak, eventually too weak so they are unable to have blood drawn; which often leads to their collapse (Interview). If they collapse, then blood transfusions may be seen as useless because the patient’s veins are too weak to support them. Also, constant blood transfusions can lead to an iron overload which is an over access of iron. Even though patients with sickle cell anemia usually suffer from iron deficiency, iron overload can be dangerous just as any other overload of minerals would. Fortunately, another treatment has been used. In an article about sickle cell disease and its relation to stroke, the authors describe an experiment in 1999 by [Russell E. Ware](http://bloodjournal.hematologylibrary.org/search?author1=Russell+E.+Ware&sortspec=date&submit=Submit),

[Sherri A. Zimmerman](http://bloodjournal.hematologylibrary.org/search?author1=Sherri+A.+Zimmerman&sortspec=date&submit=Submit) and [William H. Schultz](http://bloodjournal.hematologylibrary.org/search?author1=William+H.+Schultz&sortspec=date&submit=Submit) in which they used hydroxyurea as an alternative for blood transfusions. In the experiment,

Ware et alexamined 16 children with stroke histories who were on chronic blood transfusions for a mean of 56 months and had their transfusions stopped because of alloimmunization, autoimmune hemolysis, iron overload, or noncompliance. HU therapy and intermittent phlebotomy to remove iron were offered to these patients, beginning 2 weeks after the last transfusion. After a mean of 22 months on HU therapy, the stroke rate was 19%. Although significantly lower compared with historical controls,[17](http://bloodjournal.hematologylibrary.org/content/114/25/5117.full#ref-17) the stroke rate was almost twice as high as that observed in patients receiving transfusions in the same institution (11%). Notably, the average time to stroke in those who had the complication was only 3 to 4 months. (Nathan, David G; Verduzco, Luis A.)

With new test using hydroxyurea, sickle cell patients are able to rest their veins while receiving a more effective treatment, lasting longer than blood transfusions. However, one would now start to wonder, is there a better treatment out there that has a better effect than hydroxurea?

Due to increased cardiac output in sickle cell patients, the arteries are required to increase blood flow of deoxygenated blood. Because of the lack of oxygen in this blood, it makes the lungs work harder, which decreases the amount of exercise the sickle cell patient can endure. This increased pressure in the lungs is known as Pulmonary Hypertension, and is a commonly caused in sickle cell patients (Clarke, Melissa E. MD ) Most treatments for pulmonary hypertension include high doses of drugs such as prostacyclin. Prostacyclin has been shown to aggressively attack this disease and significantly increase quality of life, and endurance. Another option less common and for people who will not respond to the medicine is a lung transplant. Lung transplants are a good option for these patients because it reduces the amount of pressure on the lungs and no reports of pulmonary hypertension becoming worse has never been reported. Survival rates for the lung transplant have been over 50 percent, so it looks good as an alternative. However, the wait for this transplant can take years and the life expectancy after diagnosis is approximately 2.8 years in some cases (NAUSER, M.D., TRENTON D.; STITES, M.D., STEVEN W.). A sickle cell patient waiting for a lung transplant may not be worth the wait, as new damage to the lungs would occur with each sickle cell crisis they endure. Having both diseases at the same time already reduces life expectancy lower than the average sickle cell patient; therefore, drugs for treatment still stand as the most effective way for treatment.

During Cheryl’s many stays in the hospital, she often had to have aid when breathing, by using the ventilator. Her respiratory system was weak, and the constant sickle cell crises were not making it any better. She would start to complain of chest pains, which were hard to decipher between either sickle cell crisis or acute chest syndrome. Acute chest syndrome can be caused by an infection and other ways, and the symptoms may sometimes appear as symptoms for other illnesses. It is followed by high white blood cell count, but low oxygen count; severe chest pains, and fever. This can be life threatening if not treated immediately, and most treatments include blood transfusions, high doses of medicine or supplementing the patient with oxygen (WETHERS, M.D, DORIS L.)

In terms of misdiagnosis, it is very easy to mix up sickle cell with another disease because of its ability to stretch and conform into other diseases so quickly. Doctors must be able to accurately recognize the symptoms and treat the illnesses correctly otherwise, the patient will become worse. When analyzing the methods of treatment for the three complications listed above, blood transfusion is commonplace, as well as oxygen supplements. These two techniques have seemed to work; however, if continued constantly, they will start to see negative effects. One problem that prohibits scientist from finding an answer is that there is no treatment found that could stop the sickle cells from causing these complications; and this problem ties into the unanswerable question. If the life of a sickle cell were prolonged long enough for new, healthy red blood cells to be made, many of these complications could be avoided. Also, the discovery could be modified to use on other blood disorders with healthy red blood cells instead of having a specific treatment for sickled blood cells.