Sickle Cell Anemia Disease Treatment by Blood Transfusion

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ABSTRACT

Sickle cell anemia is due to single mutation subsequent in the formation of abnormal hemoglobin is called sickle hemoglobin. The abnormal hemoglobin causes alteration of red blood cells, anemia, vaso-occlusion (e.g. stroke) and dysfunction in any organ system in the body. Blood transfusion becomes the treatment and prevention of severe and persistent problems of Sickle Cell Disease. Two different types of blood cell transfusion are Simple transfusion and replace transfusion. Addition of healthy red blood cells to the patient’s body in simple transfusion. In exchange transfusions, the patient has sickle-shaped blood cells. These are exchanged with healthy ones and lowering the mediation of sickle cells without increasing blood viscosity. Change of blood is important treatment for the sickle cell disease. After every therapy the sickle cell reduces to 10 units, an involvement that replaces circulating sickle red blood cells (RBCs) with donor RBCs. The red cells are isolated from the other parts of blood. By blood transfusion, the number of normal red blood cells increases in flow, helping to reduce anemia. When normal red blood cells are transfusing and it may help supply oxygen to the body and unblock blood vessels. Blood transfusion can reduce the production of new sickle cells.

Keywords: Mutation, Transfusion, Hemoglobin, Blood viscosity.

Abbreviation

Sickle cell anemia (SCA) Sickle cell disease (SCD), Red blood cell (RBC), Delayed Hemolytic Transfusion Reaction (DHTR). Red cell exchange (RCE). Fraction cellular remaining.

Introduction

Sickle cell anemia is defined as homozygosity for sickle cell hemoglobin gene. This disease caused by missense mutation in the B-globin gene. This change occurs by a single nucleotide substitution GAG to GTG codon that codes for amino acid 6 that changes from (glutamate to valine). Red blood transfusion has an important role in the treatment of SCD in both emergency and elective setting. Sickle cell disease relates to all genotypes of the disease and homozygous state. Red blood transfusion is an essential part when acute complications takes place during sickle cell disease (SCD) in adulthood, for example, acute chest disease, priapism, splenic sequestration or stroke. Sickle cell disease patients may suffer a Delayed Hemolytic Transfusion reaction (DHTR). DHTR indications usually appear 5-15 days after complications during transfusion. Hemolysis most of the times increases piercingly during DHTR because of the devastation of both of transfused and autologous RBCs. Red blood cells (RBC) exchange, either simple handling, is a well-known treatment option for both frequent and keen hurdle of SCD. SDC patients do not breed HbA and hemolysis in the exchange of RBCs is the main aspect of DHTR. By using change exchanged hemoglobin A seems a more linked loom to analyze DHTR of keen as compared to the fall in total Hb. Treatment of severe complication is difficult because of the pathophysiological SCD. This condition is important which are correctly recognized and rapidly managed to avoid organ that acute failure. Red blood cells exchange has been mostly used for transfusion therapy as it results in lower accumulation of iron. RCE is an isovolumic transfusion that can be used to lower Hbs to a predefined level. The causes for multifactorial but some of the most common disease inherent to the erythrocytes, especially the hemoglobinopathies. The hemoglobinopathies is most common monogenic disease.
Transfusion as the Therapy for the treatment of sickle cell disease

Transfusion has been suggested for the treatment of the sickle cell anemia disease since the disease has been first described. Herrick first explained the characteristic “sickle shape” of RBCs in 1910 in the African-American dental student. After this, the transfusion medicine has been formed after World War I than the transfusion becomes the therapeutic option for the treatment of the sickle cell disease.

Acute Transfusion therapy for sickle cell disease

Acute transfusion remedy is used as the treatment of the many acute crises such as acute chest syndrome stroke and splenic sequestration (7). If the most of the organ failure occurs the exchange or partial exchange transfusion occur with some success the reverse of this is the failure of the organ.(7)

Chronic Transfusion therapy for Sickle Cell Disease

Chronic transfusion therapy has become the standard therapy for the treatment of the sickle cellular ailment from the time when the STOP1 and STOP2 display and the transfusion therapy come to be the number one and the secondary prevention for the large vessel ischemic stroke for the most risky sickness procedure in the affected person with SCA (7). Continual transfusion therapy has been used for many times to deal with the extended and persistent ache and chronic renal failure, pulmonary hypertension, and chronic acute chest syndrome. The decision whether to use red cellular exchange our simple persistent therapy depends upon the patient medical state of the patient and the available resources. Their cell exchange best constrained to the overload of the iron to a point but has a privileged on the blood deliver and continual facet results which include hyperviscosity syndrome, hemolytic transfusion reactions, extend overload and iron overload. With appropriate pre-transfusion care that consist of blood type normal screening of alloantibodies.

RBCs replace procedure

Replacement of red blood cells remedy is one of the important treatments of sickle cell mobile disorder and significantly reduces morbidity and mortality. RCE is used for blood therapy. Its replacement effects in a decrease accretion of iron. RCE is the lower evolutionary transfusion which could lesser HbS on the predefined factor(8) RCE calls for particular apparatus and experienced individual and expose to a person to have a additional red cell unit. The principle goal of exchange of RCE is the removal non-moderate or odd RBCs related with the regular suit donor red cell. The manual technique has been changed with the automated RCE finished with apheresis tool. The primary purpose of the crimson blood cell exchange(RCE) remedy is the removal of unusual Red Blood Cells (RBCs) with the ordinary healthy donor cell(9) The manual (RCE) change therapy turned into first experienced in the 1960s and 1970s and it changed into very difficult process and require an open technique. By means of mid 1980 s the guide device was exchanged with the fixed system. The predetermined structure is connected without delay with the included and programmable computers. This system enables and arranges collection and replacement based totally on the affected person scientific fact which is subsequent to the alternate technique (10). Putting the patient gender, height, weight and hematocrit data allows gathering general blood degree and the RBCs stage. Automated equipment calculate the change of red blood cell level require to get the goal.
HbS and RBCs levels. RBC chamber is used to the exchange fluid. When washer unit has used the replacement of the blood unit hematocrit level reached to the extent of the 70%. The general idea is that the final hematocrit level is needed to prevent side effects(5). An anticoagulant: The ratio of entire blood can be put to the precise ratio via the manufacturer Anticoagulant Dextrose answer A (ACD-A) and Citrate Dextrose Phosphate is used in the alternation of RCE(11)

**Growing the inlet:** ACD ratio reduces the difficulties which are associated with the citrate. The fraction cellular remaining(FCR) is the proportion of the unusual cellular left at the top of the complete procedure (12) In previous studies suggest the FCR ratio is <30% then it gives the advantageous outcome of the patient. (6)

**Benefits of Transfusion**

It gives that normal blood cell in the blood circulation. It allows more hemoglobin for the better delivery of the oxygen in the body Prevent blockage of flow in the blood vessels and decrease the need to produce new red blood cell because the transfused red blood cell lives more in the cell than the sickle cell shape(13). Chronic therapy greatly decreases the risk of health problems caused by sickle cell disease such as chronic chest syndrome. It also reduces the risk of the stroke or the stork occurrence.

**Case 1:**

Transfusion remedy leads to predominant enhancement in blood viscosity, red cell deformability, and pink cell accumulation, however there can be major rheologic and physiologic variation among simple chronic transfusion and red blood cell trade that exceed easy substitution of SS RBC with AA RBC. A number of the rheologic abnormalities are better but no longer completely corrected, which may permit odd microcirculatory drift and decreased A 28-12-month young man with SCD reachable to Methodist hospital (Omaha, NE) with serious musculoskeletal pain regularity with SCC. While the patient became admitted to the clinic, the Hb level became nine.8 g/dL, reticulocyte be counted 15%, and the ache changed into treated with medicine. On sanatorium Day 2, he changed into identified with severe chest syndrome and minor contamination due to network obtained pneumonia. By way of day 4 his Hb degree reduce to 8.6 g/dL, his and his pain growth gradually, and his coronary heart rate elevated to 155 with severe coronary heart beat, which become treated with tablets. He turned into iron and EPO on Day 6. By Day 7 his Hb level reduced to a nadir of 3.7 g/dL and he developed the heart sickness with poor blood and urine cultures. The affected person become transfused with the RBCs, however no similarity of RBCs was created, as a consequence The affected person became was given approximately 1.8 devices/day over the next 2 weeks, for complete of 27 gadgets, during which his Hb stage increased to 6.4 g/dL. This became accompanied because of an increase in free Hb from zero.1 to 1.1 g/dL. Imly arterial stress expanded from time to time as excessive as 95 mmHg. No remedy was turned into given high blood pressure. The affected person pain turned into much less, and he became discharged from Hospital Day 22, with a Hb stage of 5.7 g/(14)

**Case 2:**

A female having sickle cell anemia treated with blood transfusion remedy. When she receives her last transfusion in a month she was getting pain in some parts of body. Cefepime and vancomycin and giving in her veins as
antibiotics. After getting antibiotics and transfusion treatment a blood count test is applied and it had been seen that the rate of blood cells increases after treatment. Patient had normal anti-c and anti-k cells when they are compared with purple red blood cells. However, while, she was admitted to the hospital, DAT turned into tremendous with some antigen and C3 more attuned with heat autoimmune hemolytic anemia. Extraction becomes tremendous. The female prescribed of purple red blood cells and giving her another dose known as “prednisone” tablets two times each day for hemolysis due to autoantibodies. Hemolysis transporter non withstanding steroid treatment so broad and slight smears had been dispatched which exposed babesia contamination with a dose of parastemia 3.9% of red blood cells. The patient becomes started on quinine and a fungal derivative medicine. Each medicine was not now continued and the affected person was treated with two other medicines. The outer maximum coat became negative after approximately 1 month of remedy, however the PCR shows side effects after 3 months of treatment. But after treatment with these medicines the blood shows some germs which cause an effective reaction in the blood then this female future treated with more medicines for 2 months.(9)

**Conclusion**

Transfusion therapy leads to major enhancement in blood viscosity, red cell deformability, and red cell accumulation, but there may be major rheologic and physiologic differences between simple chronic transfusion and red blood cell exchange that exceed simple replacement of SS RBC with AA RBC. Many of the rheologic abnormalities are enhanced but not fully corrected, which may allow abnormal microcirculatory flow and decreased tissue oxygenation continue in sickle cell disease(7) RBC transfusion has long been an essential part of the management of SCD. The careful use of blood can be both lifesaving and sustain life in a variety of clinical settings. New methods may someday replace this type of therapy clinicians associated with the transfusion in the SCD.(15).

**REFERENCES**


