

Acute Transfusion for Vaso-Occlusive Crisis in Sickle Cell Disease

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Synopsis:

Patients affected by sickle cell disease suffer complications related to virtually every organ system, frequently requiring admission to the hospital. The most common manifestation of sickle cell disease and the most common cause of hospital admission for these patients is vaso-occlusive pain crisis. This results in significant morbidity for these patients including impaired quality of life secondary to pain during admission as well as missed school and work for patients and their families.

Transfusion, used either acutely or chronically, is known to be beneficial in nearly every manifestation of sickle cell disease. It is known that chronic transfusions prevent the development of VOC but the efficacy of transfusions in hospitalized patient with acute VOC has never been fully investigated.

In a pilot study at CHRCO, transfusion given to prevent acute chest syndrome reduced the length of hospitalization in VOC patients by 27% despite the fact that transfusion was given three days into hospitalization. This preliminary evidence suggests that transfusion initiated within 24 hours of presenting with VOC will result in an improvement in pain, less opioid use and a shorter length of hospitalization.

This study proposes to randomize sickle cell patients presenting with VOC to standard care or standard care in addition to a one time 10cc/kg packed red blood cell transfusion. The primary objective will be to determine if transfusion given within 24 hours of presentation for vaso-occlusive crisis will decrease the length of hospitalization in sickle cell patients. Secondary objectives will include (1) to determine if transfusion will result in an improvement in pain scores reported by patients (2) to determine if transfusion will decrease the amount of opioid use in patients hospitalized for VOC (3) to determine if transfusion will increase the amount of time before re-presentation with a new episode of VOC (4) to monitor sickle cell patients for acute and delayed complications associated with transfusion as well as development of allo-antibodies after transfusion (5) comparison of gene expression profiles pre and post transfusion to determine pathways by which transfusion provides benefit to the patient with VOC.

If shown to be efficacious, this therapy could have an enormous impact on the care for and quality of life for sickle cell patients who are frequently affected by vaso-occlusive pain episodes