

PROTOCOL #22 - Maternal Fetal Medicine, University of New Mexico

**SICKLE CELL CRISIS IN PREGNANT PATIENTS**

A. General

Sickle cell disease is an autosomal recessive disorder characterized by the presence of a predominance (usually >60%) of hemoglobin S in place of hemoglobin A as diagnosed on hemoglobin electrophoresis. In the homozygous form, "SC disease", frequent painful crises may occur as a result of hypoxia, infection, or cryptogenic causes. These crises are usually characterized by abdominal and/or bone pain as a result of tissue ischemia secondary to deformation ("sickling") and subsequent sludging of erythrocytes with occlusion of normal capillary flow. Such episodes may be exacerbated during pregnancy and may result in serious maternal sequelae and fetal jeopardy. Patients with "SC disease" may also experience similar episodes. These episodes are not seen in heterozygous individuals with "sickle trait".

B. Management

1. The goal of therapy is the relief of symptoms and achievement of a hemoglobin A concentration of >50% and assurance of fetal well being.
2. Keep the patient at bedrest, monitor vital signs and fetal heart rate, and hydrate with Ringer's lactate solution (bolus 1 L over 1 hour, then continue at 125 mL/hr) maintaining scrupulous intake and output records.
3. Provide analgesia as needed with acetaminophen, codeine, or morphine as needed.
4. Obtain an arterial blood gas or determine oxygen saturation and if evidence of hypoxia ( $pO_2 < 60$  mm Hg or  $SaO_2 < 92\%$ ) administer oxygen by nasal prongs at 3-6 L/min.
5. Partial exchange transfusion is the most important therapeutic maneuver in painful vaso-occlusive crisis in order to both reduce the concentration of Hgb S and improve  $O_2$  carrying capacity. Hematologic consultation is required and optimally requires use of an IBM 2997 Cell Separator for continuous automated erythrocytapheresis. Six units of PRBC are usually exchanged. Careful monitoring of withdrawal and return rates are critical in order to avoid overload or hypovolemia.
6. In the absence of this technology, a partial exchange transfusion may be effected with a manual protocol.
  - a. Procedure
    1. Obtain baseline Hct, quantitative Hb electrophoresis (for % HbS) and % RBC with sickling.
    2. Type and cross match for four units of fresh buffy coat poor, washed, concentrated red blood cells.
    3. Warm up cells before infusion.
    4. Infuse 500 ml Ringer's lactate in one hour.
    5. Remove 500 ml of blood from patient over a 30 minute period.
    6. Infuse two units (300-400 ml) of the red cells over one to two hours. Cells should be warmed under pressure.
    7. Repeat procedure 12 hours later.
    8. Repeat lab data (#1) 24 hours after second transfusion and every two

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weeks thereafter.

9. Repeat transfusion if Hct < 25%, HbA < 20%, or if % RBC that sickle is > 60%.

b. Equipment

1. #16 or #14 angiocath
  2. Pharmaseal K50L
  3. Extension tubing
  4. Peritoneal dialysis catheter connector
  5. 15-gauge needle
  6. 50 ml vacutainer bottle
7. Patients who are severely anemic (Hct<1 5%) should be transfused to a Hct>30%.
  8. Be vigilant for the complications of occult infection, especially of the urinary tract, congestive heart failure, and pulmonary emboli, especially in the postpartum period.

**CONSULTATION:** Twenty-four hour consultation is available by calling the Maternal Fetal Medicine service at the University of New Mexico Hospital. 1-888-866-7257.