**Sickle Cell Disease: Inpatient Care**  
4/13/2011

Most common reason for admission: Sickle Cell Disease related pain crisis
- Pathophysiology: multicytic flow obstructing aggregates form on endothelial surfaces in post-capillary venules causing downstream tissue ischemia.
- Pain= tissue damage; recurrent tissue damage leads to progressive end organ dysfunction and organ failure.
- Always believe the pain complaint: early, aggressive therapy limits tissue damage.
- Triggers: Infection/inflammation, dehydration (including from vomiting/diarrhea and intrinsic renal concentrating defect), change in weather patterns (especially exposure to cold), increased hemolysis, idiopathic.

**BE VIGILANT:**~ 30% of deaths in patients with Sickle Cell Disease occur within the first 24 hours of admission and ~60% of deaths occur within days of an admission for pain crisis.

Most common cause of death in Sickle Cell Disease (factors to consider on admission): 1) infection, 2) acute chest syndrome, 3) stroke/ICH, 4) other thrombosis, 5) acute end organ failure (kidney/liver), 6) chronic end organ failure (pulmonary hypertension, kidney/liver).

Note: Acute chest syndrome is a clinical definition based on 1) new non-atelectatic infiltrate on plain film involving at least a whole lobe, 2) fever, 3) chest pain, 4) cough/SOB/hypoxemia.

**BE CAREFUL:** Most common contributors to death/complications in Sickle Cell Disease are **IATROGENIC**
- Fluid overload (pulmonary edema ➔ hypoxemia), narcotic overdose (respiratory depression ➔ hypoxemia), delayed therapy, missed infection, transfusion complications (iron overload, alloimmunization, transfusion reactions)

**MANAGEMENT:**
- Evaluate trigger and focal pain complaints thoroughly for treatable causes
- Initial lab screen should include CBC with diff, complete metabolic panel, retic count, blood cultures, UA/culture, LDH, magnesium (Hemoglobin F% only useful to monitor hydrea response, Hemoglobin S% only if considering exchange transfusion).
- Chest XR (PA and lateral).
- IV fluids: **HYPOTONIC FLUIDS** only (1/2 NS or D5W) and supplement magnesium to upper limit of normal (2.2-2.4mg/dl) to promote RBC hydration. Avoid/monitor for pulmonary edema.
- Incentive spirometry q hour while awake
- IV narcotics are mainstay of pain control therapy, anti-inflammatories during first 48 hours usually recommended
- Include macrolide (azithromycin) to antibiotic regimen if pneumonia suspected (atypical bugs especially mycoplasma and chlamydia are common in SCD)
- Continue hydrea and folate during admission at outpatient doses
- Transfusion:
  o NOT indicated for simple pain crisis.
  o Transfuse for symptomatic anemia or if PRCA suspected (inappropriately low retic count with worsening anemia).
  o NEVER transfuse to hematocrit over 28% (hyperviscosity complications including stroke).
  o Target end transfusion hematocrit depends on the patient and clinic scenario.
  o Blood products should be leukoreduced and antigen matched for ABO, Rh D, Rh C/E, and Kell (write "FOR SICKLE CELL PATIENT" on blood order form to trigger PSBC protocol for this).

Patients are instructed to remain active/ambulatory during pain crises: **Never mistake activity for lack of a real pain complaint.**