Sickle Cell: Common ER presentation

What causes sickling? Low O2, low pH, dehydration, cold temperature

1. **Acute Chest**
   1. Epidemiology: highest incidence in 2 to 5 year olds
   2. Definition: new pulmonary infiltrate on CXR with one or more of the following: fever, tachypnea, dyspnea, hypoxia, and chest pain
   3. Causes: infectious (chlamydia pneumonia, viral infection, mycoplasma), pulmonary infarction, fat embolism, or any other type of regional alveolar hypoxia (atelectasis from VOC of ribs, pulmonary edema, bronchospasm, etc)
   4. Treatment: O2, antibiotics (CTX + azithromycin), bronchodilators (albuterol, inhaled corticosteroid for “pulmonary sick plan”), pain management, IV fluids (0.75x maintenance)
2. **Pain crisis: vaso-occlusive crisis**
   1. Epidemiology: infancy until older age🡪 childhood to mid 20s
   2. Pathophysiology: bone marrow ischemia with resultant infarction, triggered by infection, emotional stress, exposure (cold, wind, high altitude)
   3. Presentation: point tenderness or swelling/erythema if dactylitis
   4. Who gets admitted? Requiring more than 3 doses of opioid without improvement
   5. Treatment: pain management (opioid + Toradol), IV fluids (1x mIVF)

*Dactylitis:* Specific type of VOC that occurs in young children and infants (erythema, swelling, and tenderness, age 1-2 years)

1. **Splenic sequestration**
   1. Definition: rapid enlargement of spleen with resultant trapping of blood
   2. Presentation: splenomegaly, Hgb below baseline, thrombocytopenia
   3. Epidemiology: SS disease= 30% of children by 5 years of age w/ most episodes before 2 years of age; SC disease >10 years or older
   4. Treatment: Splenectomy after second or third sequestration episode, serial exam, if enlarging stat CBC🡪 transfuse then PICU?
2. **Sickle cell with fever (>38.3C)**
   1. ER workup: CBC, blood culture, CTX, bolus
   2. Who gets admitted? High risk features (WBC>30K or <5, fever >40C, “ill-appearing”), young children, inability to return promptly for recurrent fever
   3. Treatment: CTX, 1x mIVF, anti-pyretic

*Chronic preventative treatment*: hydroxyurea (increases fetal hemoglobin) so prevents sickling and thus decreases risk for ACS, transfusion, and VOC