**EM Basic- Sickle Cell Anemia**

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**Background**

Acute pain crisis is the most common presentation

Remember to rule out life threatening diagnosis

Do not anchor on typical pain crisis if something doesn’t feel right

Hgb SS-most common/severe, Hgb SC-less severe/similar complications

**History**

**HPI**

OPQRST-chest pain/sob, fever/chills, back pain, abd pain, cough

Does this feel typical of pain crisis? Is there anything new/different?

Baseline hemoglobin level? Last transfusion?

**Medications** – pain meds-what regimen they’re on and what are they taking at home

Hydroxyurea, folic acid

Prophylactic abx- penicillin in peds

Immunizations-pneumococcus, influenza, etc.

**PMH** – hx of CVA, tobacco, alcohol, hx of acute chest syndrome/PE, ACS

**Social** – illicit drug use

**PEARL-** can still have normal vital signs in the setting of acute pain

**Physical Exam**

**Vitals** – tachycardia, tachypnea, hypotension, T > 38 C

**General** – sick or not sick, toxic appearing

**Neuro-**as needed if focal weakness, AMS

**Lung**-rales, wheezing, retractions/resp distress

**Abd**-peritonitis, hepato-splenomegaly

**MSK**-bony tenderness, septic arthritis, osteomyelitis

**Skin**-cellulitis, abscess, infection

**PEARL**- most important part of exam is looking for source of infection

**Workup**

**CBC**-baseline Hgb

**Reticulocyte count**: hemolysis, aplastic crisis (low)

**BMP-** renal function

**LFTs**- transaminitis, elevated alk phos, bilirubin (hepatic crisis, AIC (acute intrahepatic cholestasis)

**Lactate level**-as needed for sepsis

**Type/screen/crossmatch**-if anticipating transfusion

**ABG/VBG-**as needed for resp distress

**EKG**-STEMI, ischemia, signs of PE

**PEARL**- labs generally not helpful in uncomplicated pain crisis

**Imaging**

**CXR**- consolidation, pulm edema, atelectasis

**Head CT-** signs of CVA

**CTA chest**- PE, fat embolism

**Differential Diagnosis**

**Stroke/CVA-**focal weakness, AMS, slurred speech

**Acute chest syndrome**-fever, cough, sob, resp distress, hypoxia, new findings on CXR

**Vaso-occlusive crisis**-back, joint, chest wall pain, dull/achy (diagnosis of exclusion)

**Aplastic crisis**-lethargy, weakness, viral syndrome, low retic count, acute drop in Hgb

**Splenic sequestration**- LUQ pain, splenomegaly, hypotension, pallor, shock, low hemoglobin. More common in peds

**Hepatic crisis/AIC**- RUQ pain, hepatomegaly, shock, lethargy, hypotension, elevated LFTs, bilirubin

**PE**-chest pain/SOB, tachycardia, hypotension (massive)

**ACS**-chest pain, sob, weakness, n/v

**Sepsis/infection**- be on the lookout for meningitis/encephalitis, cellulitis, abscess, septic arthritis, osteomyelitis

**PEARL-** have low threshold for blood cultures, admission in fever without an obvious source

**Management**

**ABCs** – intubation, O2, bipap, IVFs if hypotensive, observe resp function closely

**Acute vaso-occlusive crisis**

-pain management, 6-8mg morphine, 0.5-1mg hydromorphone IV, every 15-30min until adequate pain control. Can add ketorolac, ketamine.

-oxygen only if they are hypoxic, maintenance IVFs, oral rehydration if tolerating PO, only bolus if hypotensive, avoid lots of NS

**PEARL**- do not transfuse uncomplicated acute pain crisis

**Acute chest syndrome**

-supportive care, support respiratory function (O2, bipap, intubation), IVFs (careful to not over resuscitate)

-ABX (CAP coverage), simple/exchange transfusion if they do not improve

**Splenic sequestration/hepatic crisis**- supportive care, volume resuscitation, simple/exchange transfusion

**Aplastic crisis**- supportive care, simple transfusion if severely symptomatic

**Stroke/CVA**- exchange transfusion

**Sepsis**- blood cultures, broad spectrum abx, IVFs as needed, supportive care, close monitoring

**Disposition**

**Discharge:** uncomplicated pain crisis if tolerating PO and adequate pain control is achieved. Ensure adequate follow up with hematologist/sickle cell clinic.

**Floor/progressive care**: sepsis if hemodynamically stable, aplastic crisis. Intractable pain in the setting of acute pain crisis.

**ICU:** acute chest syndrome, splenic sequestration, hepatic crisis, septic shock, massive PE. Vasopressor requirement, significant resp distress requiring bipap/intubation.

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