

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

Workup

CBC-baseline Hgb

Reticulocyte count: hemolysis, aplastic crisis (low)

BMP- renal function

LFTs- transaminitis, elevated alk phos, bilirubin (hepatic crisis, AIC (ac 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 exclusion)

Aplastic crisis-lethargy, weakness, viral syndrome, low retic count, ac 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, cellul abscess, septic arthritis, osteomyelitis

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

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/sic 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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