Acute Management of Sickle Cell Crises

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SICKLE CELL DISEASE

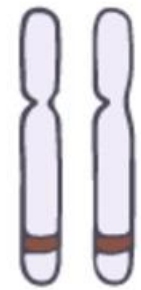
SICKLE CELL ANEMIA

easily DESTROYED

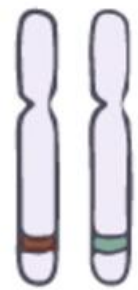
AUTOSOMAL RECESSIVE DISORDER

HEMOGLOBIN GENE

ABNORMAL ADULT HEMOGLOBIN
HEMOGLOBIN S (HbS)



MUTATION



NORMAL HEMOGLOBIN A GENE (HbA)

SICKLE TRAIT

Burden of Sickle Cell Disease (SCD)

25000/year

7% of all tests positive

15%

< 5y mortality

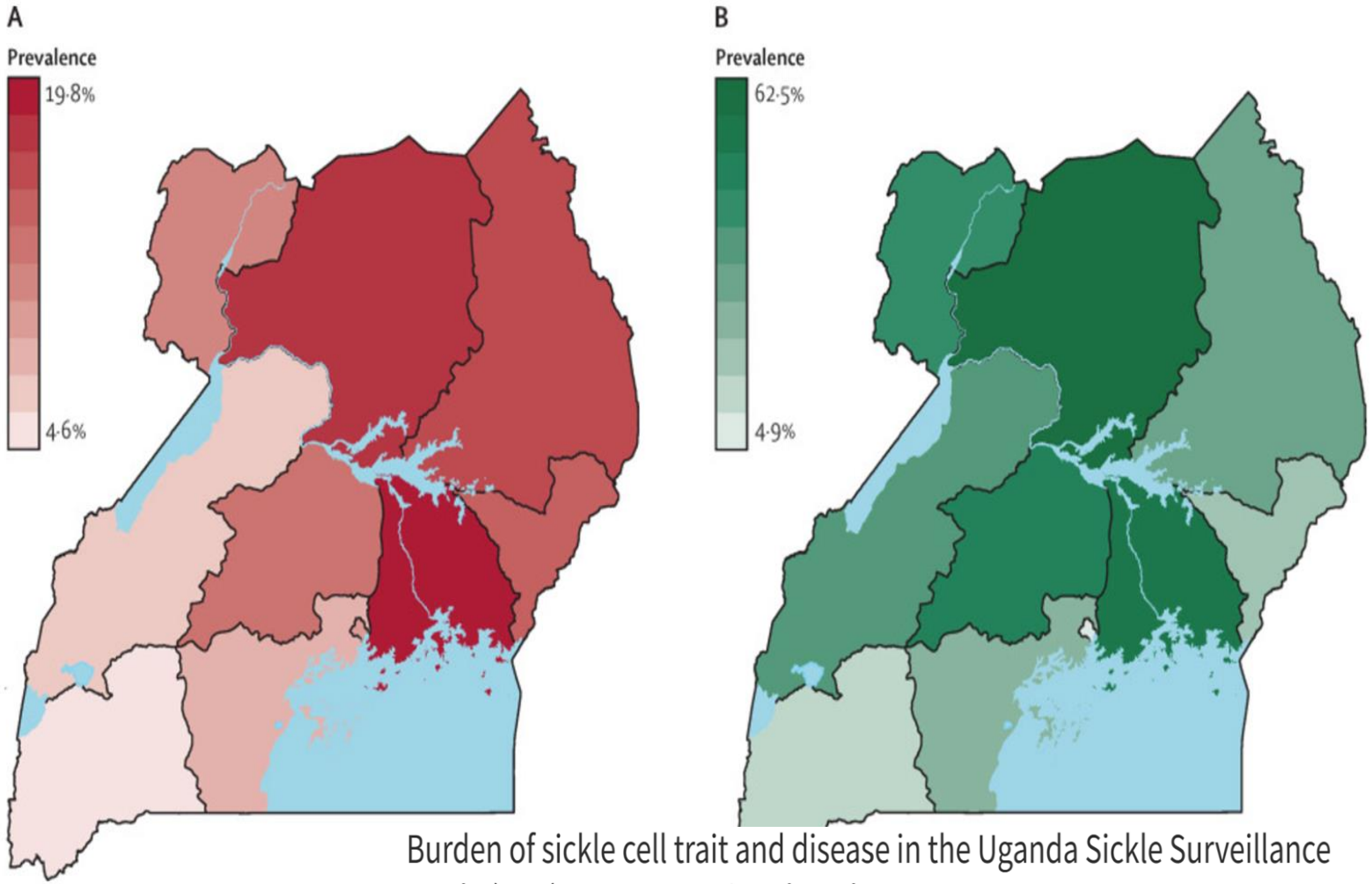


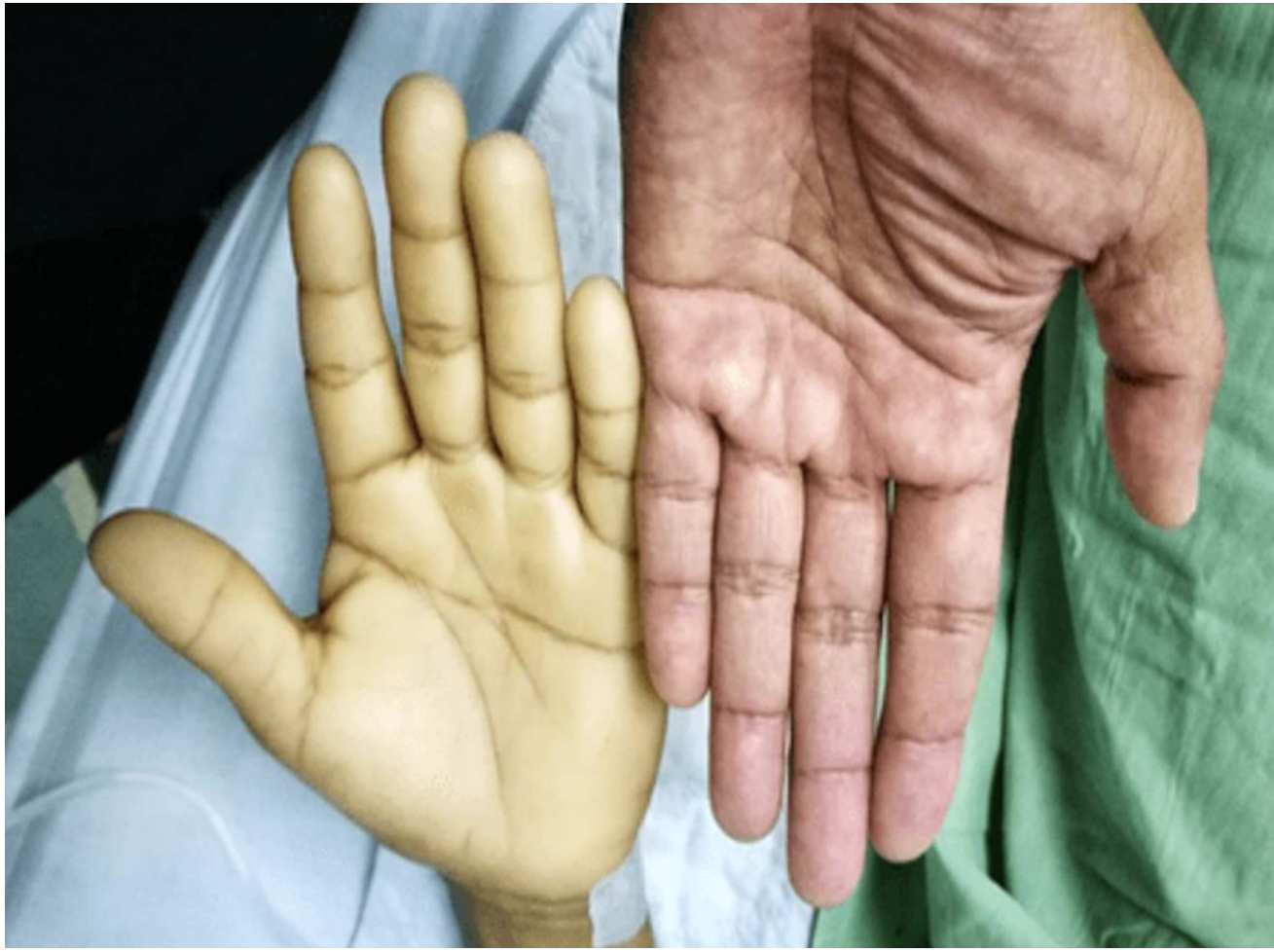
Figure 2 Prevalence of sickle cell t

Burden of sickle cell trait and disease in the Uganda Sickle Surveillance Study (US3): a cross-sectional study

Grace Ndeezi, MBChB PhD [a] · Charles Kiyaga, MSc MPhil [b] · Arielle G Hernandez, BA [c] · Deogratias Munube, MBChB [d] · Thad A Howard, MS [c] · Isaac Ssewanyana, MSc [b] · et al. Show more

Acute Management in the ED

- ABCDE Assessment:
- If actively bleeding, stop the bleeding, pack/tourniquet/TXA
- Trigger for interventions = clinical signs and symptoms



Sickle Cell Crises

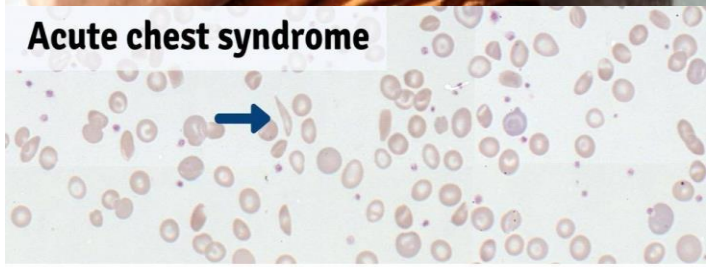
- **Acute vaso-occlusive:**
  - **Acute Pain, Acute chest, stroke**
- **Splenic sequestration**
- **Aplastic crisis**
- **Hyper-Hemolysis**

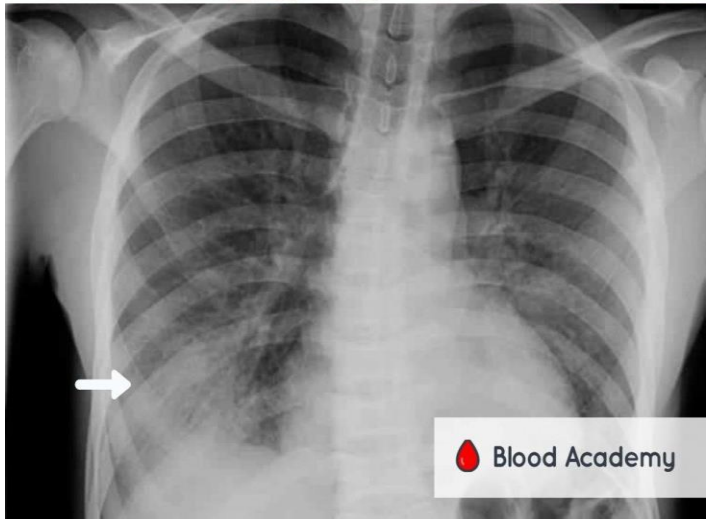
Hx in Sickle Cell Pain Crisis

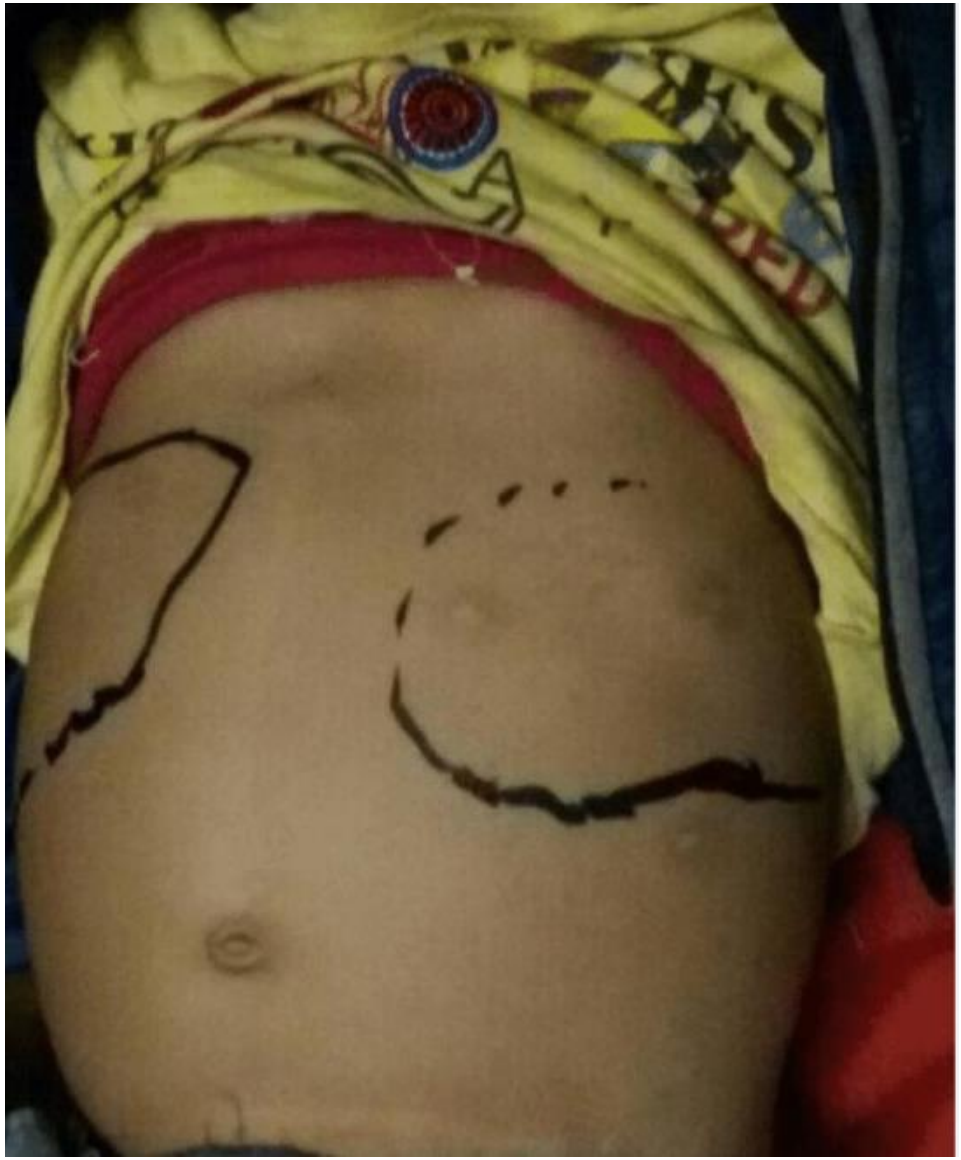
Relevant Questions:

- What sickle cell complication has the patient had in the past?
- How often does the patient have pain, and how often does the patient come to the Ed for pain?
- Has the patient ever required multiple transfusion?
- Is the patient currently taking analgesia, antibiotics or hydroxyurea?
- What is the patient’s baseline hemoglobin level?

Physical exam in Sickle Cell Pain Crisis







Management of Sickle Cell Pain Crisis

- Analgesia (iv morphine 0.1-0.2mg/kg per dose child, adult 5-10mg per dose)
- ABCDE
- Oxygen
- Hydration
- Analgesia

Acute Chest Syndrome

- Most common cause of death in SCD with fatality rate

- Classic triad; fever, hypoxia and pulmonary infiltrate

Management of Acute Chest Syndrome

- Oxygen supplementation maintain SPO2 > 95%, NIV prn
- Red cell transfusion or exchange transfusion
- Initiate broad spectrum empiric antibiotics
- Avoid fluid overload

Govt to slash price of sickle cell drug by half

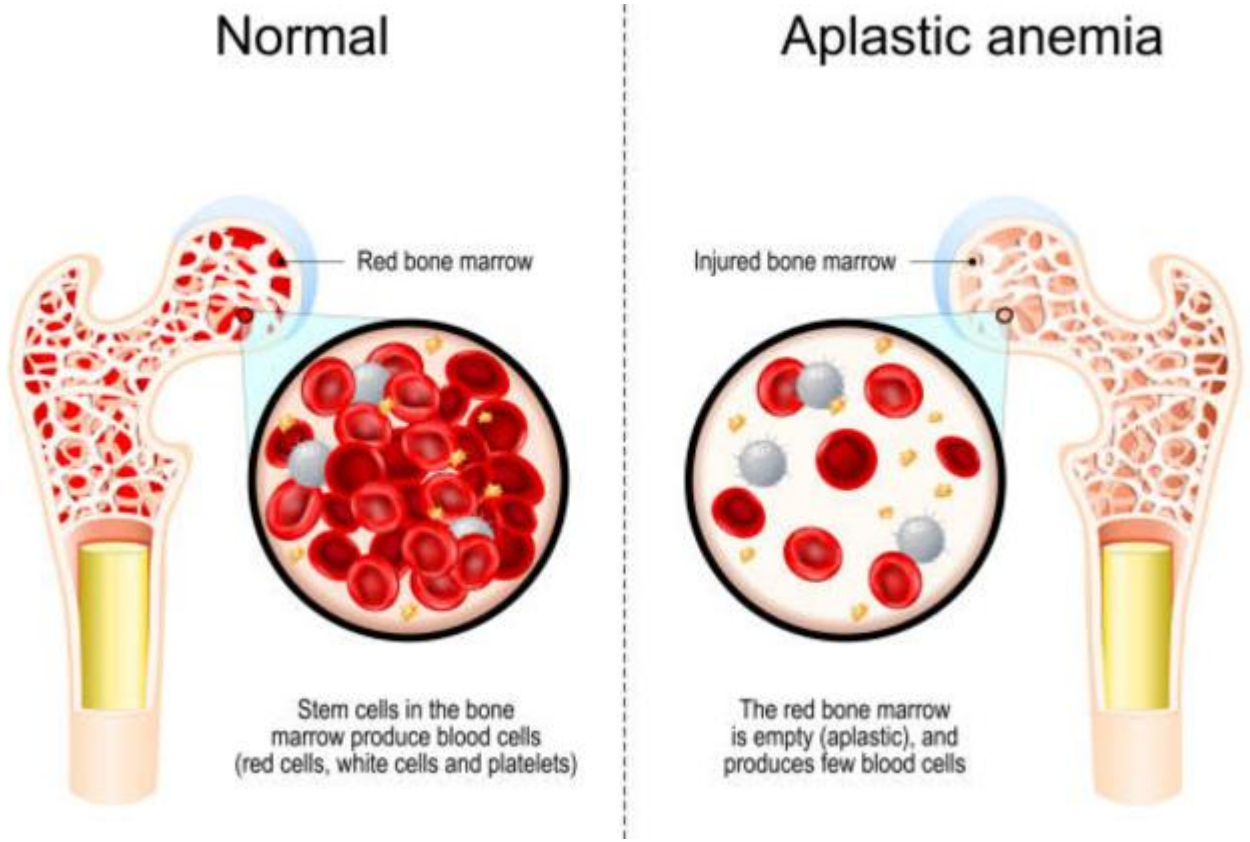
Tuesday, June 21, 2022

Hydroxyurea drugs will now be sold at about Shs700 per tablet up from Shs1,700. PHOTO/COURTESY

Aplastic Crisis

- Is a life threatening condition that can be elusive

  - ➢ Usually a viral prodrome
  - ➢ Acute severe drop in serum hemoglobin
  - ➢ Often hemodynamically compromised in shock at presentation
  - ➢ Reticulocytes count dramatically decreased or even zero



Management of Aplastic Crisis

- Immediate red cell transfusion
- Note; DDx (paediatric) = **sequestration syndrome**
- Look for tender palpable organomegaly, high indirect bilirubin, ALT and LDH and high reticulocyte count.

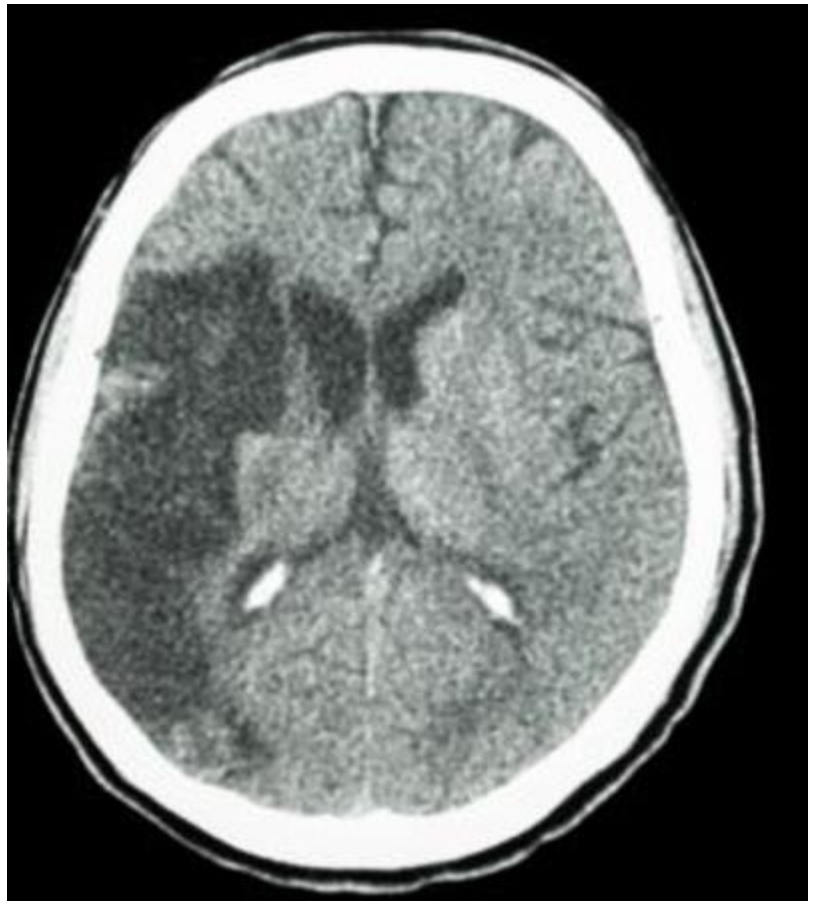
Management of Splenic Sequestration

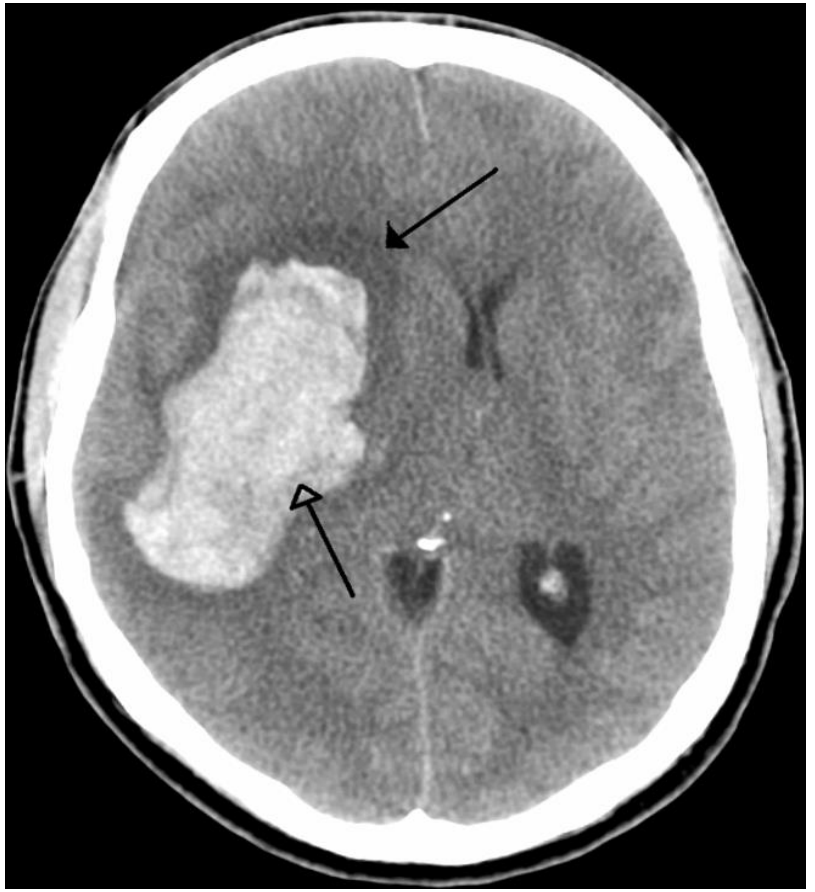
- Red cell transfusion
- Splenectomy
- Admitted to a monitored

Management of Hyper-Hemolysis

- Assess for jaundice, tea colored urine and severe anemia
- Red blood cell Transfusion

Sickle Cell Disease and Stroke





Disposition Plan for Sickle Cell Crises

Depends on:

- Severity of patients presentation
- Need for intervention from other specialties like:
- Hematology consult, orthopedic, stroke/ neuro team and respiratory specialist (ventilation)

Future Directions

- Stem cell transplant
- only available curative therapy for SCD
- Not suitable for everyone = significant toxicities associated with the procedure.
- Currently indicated in persons with severe SCD with complications

Take Home Points for ED Management

In Conclusion:

- Give oxygen only to those that need it saturation<95%
- Judicious fluid administration
- Pain management with 15-30 minutes with opiates, NSAIDs and ketamine for opiate sparing drug
- Use NSAIDS sparingly
- Reserve supplemental oxygen for patients who are hypoxic and fluid bolus for hypovolemic

Thank you All!