CASE OF A 14 MONTH OLD
WITH SICKLE CELL AND
ACUTE SPLENIC
SEQUESTRATION
Objectives

- Case presentation
- Brief overview of Sickle cell disease
- Identification and management of acute splenic sequestration
- Management of severe anemia
- Identify and manage DIC
CASE PRESENTATION
HPI

- 14 month old female presented to an outside hospital after “seizure like activity”. Patient was brought in by parents and said that she made a strange face then was stiff and making jerking motions in dad’s arms and then became limp.

- Parents stated that she had had a fever of 101 that day with vomiting, rhinorrhea and decreased PO intake
History

- PMHx: HbSS (baseline Hgb 7), recently treated for scabies
- Surgical Hx: None
- Meds: Folic acid 1mg daily, Pen VK 125mg BID
- Allergies: Augmentin, causes rash
- FamHx: siblings with sickle cell trait
Outside hospital exam

- Records indicate that she arrived unresponsive, pale, tachypneic with agonal respirations and rigid abdomen
- Vitals on admission: HR 168, RR 64, Temp 98.1
- BP throughout her time at the outside hospital 97-109/32-38
OSH initial labs

- VBG: pH 6.5, pCO2 22.1, pO2 51.7, HCO3 1.7
- WBC 34.5, Hgb 1.5, Hct 4.6, plt 57K
- Na 148, K 7.7, Cl 104, CO2 <5, BUN 18, Cr 1.3, glucose 187, Ca 9.7
She was given a 20cc/kg bolus, intubated and placed on a vent,
Transfused **2 units** of PRBCs (approx 50ml/kg over 2 hours), and given a dose of solumedrol.
Given kayexalate, albuterol, calcium chloride, insulin/D25 for the hyperkalemia.
Was also given Rocephin 50mg/kg
Patient promptly transferred to CHNOLA PICU
Arrival to CHNOLA PICU

- **Physical exam**
  - Vitals: BP **133/62**, pulse **176**, RR **67**, pulse ox 95% on FiO2 100%, temp **102.4**, weight 10.1 kg
  - Gen: agitated and combative
  - HEENT: intubated, **ETT with pink secretions, NGT with dark/frank blood**, TM clear
  - CV: **tachycardia**
  - Resp: coarse breath sounds bilaterally with **inspiratory stridor due to air leak**, + retractions
  - Abd: absent bowel sounds, **spleen palpable with tip felt in pelvis, hepatomegaly**
  - Ext: right hand swelling, dark purple in color, **cap refill >3-4 secs**, pulses 2+
CHNOLA admission labs

- Hgb 17.9
- Hct 52
- Platelets 22K
- PT 22
- INR 1.98
- PTT 44
- Fibrinogen 141
- D. dimer >20
- pH 7.16
- pCO2 46
- pO2 45
- Na 144

- K 4.1
- Cl 113
- CO2 20
- BUN 25
- Cr 0.4
- Glucose 279
- Ca 7.5
- Phos 4.9
- Mag 2.2
- AST >1000
- ALT 270
- Tbili 1.8
Labs con’t

- Viral panel: +rhino/enterovirus
- UA
  - Color brown
  - SG 1.025
  - pH 6
  - LE negative
  - + nitrite
  - 1+ protein
  - 3+ blood
  - 1+ bili
  - 3+ bacteria
  - 0-2 granular casts

- BCx: pending
- UCx: pending
Early hospital course

- Patient presented in DIC and was transfused 150ml (15cc/kg) of FFP and then 100ml of platelets (10cc/kg) with a goal platelet count of 50K
- She was started on versed, fentanyl and vecuronium for sedation and to chemically paralyze
Also started on Rocephin (75mg/kg) and Vancomycin (15mg/kg/dose) for suspected sepsis

Once stable she was sent for CT of head, chest, abdomen and pelvis
Head CT

- Normal head CT scan of the brain.
- Inflammatory changes of the paranasal sinuses and mastoids.
CT chest/abd/pelvis

- Severely enlarged hypoperfused spleen.
  Hepatomegaly with small ascites.
  Pulmonary edema with small bilateral pleural effusions. Low attenuation fullness at the root of the mesentery likely representing edema.
Echo

Sickle Cell Disease overview
Sickle Cell Disease

- Autosomal recessive inheritance
- Glutamic acid is substituted for valine which allows for the polymerization of hemoglobin when deoxygenated
- Sickled RBCs have a life span of about 20 days and are more rigid than normal RBCs
The anemia seen with SCD is caused by the destruction of the RBCs

- Degree of anemia varies patient to patient
- There is increased bone marrow production of RBCs but it is unable to keep up with the hemolysis

Vaso-occlusion is a result of the sickled cells blocking blood flow to the tissue as they are unable to move in the vessels
Genotypes

- Sickle cell anemia (Hb SS) 65%
- Sickle Hb SC 25%
- Sickle S beta plus (Sβ⁺ thalassemia) 8%
- Sickle beta zero (Sβ° thalassemia) 2%
Health Maintenance

- Mandated testing in all 50 states and Washington DC on the newborn screen
- PCN prophylaxis should begin no later than 2 months of age
- Immunizations: routine except that these children should receive 23 valent PPSV at 2 and 5 years of age
SCD emergencies...

- Fever
- Pain crisis
- Acute chest syndrome
- Acute splenic sequestration
- Stroke
Acute splenic sequestration crisis (ASSC)
Some basics

- Vaso-occlusion within the spleen and pooling of RBCs causes a drastic fall in Hgb with persistent reticulocytosis and enlarging spleen
- Associated with 10-15% mortality
- Recurrent in about 50% of survivors
Characterized by:

- Splenic enlargement
- Drop in Hgb of at least 2 grams from baseline
- Thrombocytopenia
- Reticulocytosis
Signs and symptoms

- Weakness
- Pallor
- Tachycardia
- Tachypnea
- Abdominal swelling and tenderness
Parents are instructed to examine their child’s abdomen to help catch an enlarging spleen early.

Most commonly seen in infancy and early childhood in HbSS.

Those with SC disease tend to have sequestration at an older age:
- Most frequently seen in children from 2-17 years old.
- Overall incidence about 5%.
Most drastic complication is hypovolemic shock

- Cardiovascular collapse and death can occur within 30 minutes
Acute Treatment

- IV fluids to maintain the vascular volume
- Blood transfusion to treat anemia and allow release of pooled RBCs
Chronic Treatment

- Frequent transfusions every few months to maintain Hgb close to 10
- Splenectomy if indicated, after age 2
  - Should also receive menactra and PSSV23 prior to splenectomy
Management of severe anemia
Severe anemia

At the outside hospital she presented with a Hgb of 1.5 due to splenic sequestration and was transfused 500ml of blood over 2 hours.

- Her normal entire blood volume is about 600cc
In general, guidelines support using the gram of hemoglobin as the amount per cc for transfusion when Hgb is this low.
This volume of blood likely led to her volume overload as her Hgb was 17.9 on admission

The patient actually required phlebotomy of 150cc, with a goal to reduce Hgb to 10
Management of Disseminated intravascular coagulation (DIC)
Our patient

- She presented with
  - PT 22
  - PTT 44
  - INR 1.98
  - Fibrinogen 141
  - D. dimer >20
Some basics

- Systemic activation of pathways leading to and regulating coagulation, which can cause the production of fibrin clots that may cause organ failure with simultaneous consumption of platelets and coagulation factors that may result in bleeding.
Causes

- May be caused by a wide array of clinical disorders including sepsis, trauma, liver disease, vascular anomalies, malignancy
Pathway

- Derangement of the fibrinolytic system contributes to intravascular clot formation.
- Accelerated fibrinolysis may also cause severe bleeding.
Presentation

- Hemorrhage
- Diffuse/localized thrombosis
- AMS
- Hypotension
- Tachycardia
- Friction rub
- ARDS
- Hematemesis
- Azotemia/renal failure
- Acidosis
- Petechiae, purpura
- Wound bleeding
Diagnosis

- Thrombocytopenia
- Elevated FDP
- Increased clotting time (PT and PTT)
- Low fibrinogen
  - ISTH scoring system for overt DIC
## DIC scoring system

<table>
<thead>
<tr>
<th>Laboratory Test</th>
<th>Result</th>
<th>Score</th>
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<tbody>
<tr>
<td>Platelet count (cells/uL)</td>
<td>&gt;100,000</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>50,000 – 100,000</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>&lt;50,000</td>
<td>2</td>
</tr>
<tr>
<td>Increase in fibrinogen and fibrin-related markers (eg, FDP’s)</td>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Moderately Increased</td>
<td>2</td>
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<tr>
<td></td>
<td>Strongly increased</td>
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<tr>
<td>Prolonged prothrombin time</td>
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<tr>
<td></td>
<td>3-5.9</td>
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<tr>
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<td>&gt;6</td>
<td>2</td>
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<td>Fibrinogen</td>
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</tr>
<tr>
<td></td>
<td>&lt;1 g/dL</td>
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Treatment

- Key to treatment is treating the underlying cause
- Transfusion of blood products should not be based on lab values alone
Our patient’s DIC score was 6 on admission

She received FFP on arrival and also received vitamin K 3mg daily for 3 days

She also required multiple platelet transfusions during her hospital course to maintain her count >50K
Other complications of Sickle cell in our patient
Our patient

- On hospital day 3, a repeat head CT was done that showed multifocal areas of edema that could be related to infection or infarction.
- On hospital day 4, she was noted to have seizure like activity and was loaded with keppra and then started on a maintenance dose.
- STAT head CT and MRI were done at this time.
Head CT

- Improved multifocal areas of edema, compared to the head CT from 1 day prior. Inflammatory fluid in middle ears and mastoids
MRI

- Restricted diffusion in the frontal and parietal occipital regions bilaterally without associated spin echo abnormality or abnormal contrast enhancement. Findings which could be related to hypoxia/ischemia, hypoglycemia and/or status epilepticus.
EEG

- Diffuse severe background slowing with loss of regional differentiation. Abnormal sleep forms with loss of K complexes, sleep spindle asymmetry with relatively less well sustained spindles over the left central head region.
- These findings are consistent with diffuse bihemispheric cerebral dysfunction as well as suggestive of more pronounced dysfunction over the left parasagittal area.
Stroke

- One of the leading causes of death in both children and adults with SCD
- More common in HbSS
- Type of stroke varied with age
  - More likely to be ischemic in younger patients
- “Silent infarcts”
Risk factors

- Higher cerebral blood flow in younger children
- Sludging and occlusion of blood vessels that causes ischemia
- Chronic anemia that may reduce cerebrovascular reserve
- Increased adherence of sickle cells to endothelium that may cause further endothelial injury
- Hypercoagulable state
Treatment

- Chronic transfusions to maintain sickled hemoglobin 30% or lower
  - Stroke Prevention Trial in SCD (STOP trial)
    - Iron overload
    - Alloimmunization
Our patient’s hospital course
Extubated after 9 days, had some post extubation stridor that resolved with decadron and weaned to room air

Treated with both vancomycin and claforan at meningtic doses for full 7 days, all cultures were no growth during stay. She was also placed on acyclovir for concern for HSV but was stopped when the PCR came back negative

DIC resolved prior to transfer to floor from PICU
After her seizure on hospital day 4, keppra was added. She did not have any further seizure activity while in patient and was discharged home to continue the keppra.

She had no lasting neurological deficits from her global hypoxic injury and did not have a true stroke.

She was continued on her home medications of Pen VK and folic acid.
References

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- Velez, Maria, MD. Heme-onc Emergencies. [http://www.medschool.lsuhsc.edu/pediatrics/residents/docs/Emergencies%20PHO%20for%20Interns%202011.pptx](http://www.medschool.lsuhsc.edu/pediatrics/residents/docs/Emergencies%20PHO%20for%20Interns%202011.pptx)
- CDC: sickle cell disease date and statistics: [http://www.cdc.gov/ncbddd/sicklecell/data.html](http://www.cdc.gov/ncbddd/sicklecell/data.html)
- Management of Sickle Cell Disease, 4th ed. Chapter 18: Acute splenic sequestration