Managing Pediatric Sickle Cell Patient in the Emergency Room or Urgent Care Setting

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Disclosures

- No disclosures

Learning Objectives

- List complications that occur in the pediatric sickle cell population.
- Differentiate between complications requiring hospital admission and those that can be managed outpatient.
- Discuss diagnostic and treatment plans for common pediatric sickle cell complications.

Sickle Cell Disease

- Brief overview
  - Autosomal recessive
  - Substitution of valine for glutamic acid on beta globin chain
  - Result: Sickle cells more fragile, log jam effect
  - Multiple organs affected
  - Dehydration, exposure to extreme temps, high altitude makes worse

- Types
  - Hb SS
  - Sβ⁰thalassemia
  - S↠thalassemia
  - Hb SC

- Universal Newborn Screening in U.S.
  - Diagnosis is known
  - Hemooglobin Electrophoresis

Typical laboratory findings

<table>
<thead>
<tr>
<th>Sickle Cell Disease</th>
<th>Hgb (%)</th>
<th>Hct (%)</th>
<th>MCH (pg)</th>
<th>MCV (fL)</th>
<th>MCHC (g/L)</th>
<th>RDW (fL)</th>
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<tbody>
<tr>
<td>SS</td>
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<td>31</td>
<td>35.8</td>
<td>76.8</td>
<td>0.39</td>
<td>15.8</td>
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<td>29.2</td>
<td>69.3</td>
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<td>28</td>
<td>30.2</td>
<td>69.3</td>
<td>0.39</td>
<td>15.8</td>
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<th>Sickle Cell Trait</th>
<th>Hgb (%)</th>
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<th>MCV (fL)</th>
<th>MCHC (g/L)</th>
<th>RDW (fL)</th>
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<td>36.1</td>
<td>80.2</td>
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Triage/When to admit

- Triage
  - Emergency Severity Index (ESI) version 4
    - Level 2
      - Very high priority
      - Rapid placement facilitated

- When to admit
  - Acute Chest Syndrome
  - Uncontrolled pain
  - Fever
  - Respiratory distress
  - Unstable or critical condition
Indications for transfusion

- Prior to surgery with general anesthesia
  - transfuse to a hemoglobin of 10 g/dl
- Acute chest syndrome
  - Mild-Simple transfusion
  - Severe-Exchange transfusion
- Acute splenic sequestration with anemia
- Stroke or critical transcranial dopler reading
- Hepatic sequestration
- Aplastic crisis
- Drop in baseline hemoglobin with symptoms

Transfusion Guidelines

- CBC, reticulocyte count, Hgb electrophoresis
- PRBC units matched for C, E, and K antigens
- Type and screen
- Serum ferritin periodic
- LFT's Periodic
- Avoid transfusing over 10 g/dl
- Goal to maintain HbS level at or below 30%
- 10 ml/kg is unless splenic sequestration then give less
- Iron chelation for elevated serum ferritin levels

Pediatric Sickle Cell

- How they present
  - Pain
    - Chest
    - Arms/legs/hips/back
    - Abdomen
    - Head
    - Hands/feet
    - Face
  - Fever
  - Fatigue/SIR
  - Respiratory distress
  - Lethargy
  - Jaundice
  - Vision disturbances
  - Slurred speech/numbness, tingling/weakness

Vaso Oclusive Crisis

- Diagnosis of exclusion
- Pain management
- Pain management similar
  - Vaso Oclusive Crisis
  - Avascular Necrosis
  - Osteomyelitis
  - Dactylitis
  - Priapism
  - Acute Chest syndrome
  - Splenic Sequestration
  - Gallstones

Ideal Pain Management

- Pain contracts
  - Patients maintain pain diary
    - Pain is rated mild, moderate, severe with location
    - Home treatment is documented
    - Diary is brought to ER and all clinic visits
  - Must take narcotics as prescribed
  - Only one prescriber
  - Bring all discharge summaries to clinic, mention all urgent care, ER visits
  - Bring pain medication bottles to clinic

Pain Management

- Pain Management
  - Hydration
  - Physical Therapy
    - Warm compress
    - TENS unit
    - Whirlpool bath
    - Exercise
  - Psychologic
    - Child life
    - Relaxation
    - Distraction
Pharmacologic Pain Management

- Initiate within 30 min of triage or 60 min of registration
- No Meperidine (Demerol)-CNS toxicity
  - Mild Pain
    - IV
    - Moderate-Severe pain
      - IV pain medications around the clock
  - Severe pain
    - PCA pump-start in ER
      - As young as 7-8 yrs old depending on maturity

PCA-Opioids

<table>
<thead>
<tr>
<th>Severe pain</th>
<th>Morphine</th>
<th>Hydromorphone (Dilaudid)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basal rate (mg/kg/hour)</td>
<td>0.02-0.04</td>
<td>0.002-0.007</td>
</tr>
<tr>
<td>PCA dose (mg/kg)</td>
<td>0.015</td>
<td>0.0025</td>
</tr>
<tr>
<td>Lockout period (min)</td>
<td>30 min</td>
<td>30 min</td>
</tr>
<tr>
<td>Relax dose (mg/kg)</td>
<td>0.05</td>
<td>0.05</td>
</tr>
</tbody>
</table>

IV Pain medications

- Moderate-Severe pain
  - Q 2-3 hrs IV ATC
    - Morphine 0.1-0.15 mg/kg/dose
    - Dilaudid 0.015-0.02 mg/kg/dose
    - Fentanyl (hepatic or renal impairment) 2mcg/kg/dose
    - Ketorlac 0.5 mg/kg/dose IV (max 30 mg/dose)

PO pain medications

- Mild-Moderate pain
  - Ibuprofen
    - 10mg/kg/dose
  - Oxycodone
    - 6 yrs: 0.15 mg/kg up to 2.5 mg
    - 6-12 yrs: 0.2 mg/kg up to 5 mg
    - >12 yrs: 0.2 mg/kg up to 10 mg
  - Hydromorphone (Dilaudid)
    - 0.05-0.26 mg/kg/dose PO q1-4h
    - Adult dose: 2-4mg
  - MS contin-sustained release
    - 0.3-0.6mg/kg/dose PO q12h
  - MSIR-immediate release
    - 0.2-0.5 mg/kg/dose PO q4-8h
  - Methadone
    - 0.2mg/kg/dose PO q4-6h Max 60 mg/dose
  - Codine
    - 0.5-2mg/kg/dose PO q6h Max 60 mg/dose

Pain management

- Hydroxyurea guidelines
  - Not appropriate to initiate for acute pain
  - Offered to all children with sickle cell > 9mo age for chronic pain
  - Initial starting dose 20 mg/kg/day
  - Dose escalation 5mg/kg every 8 weeks as tolerated unless target ANC reached or toxicity occurs. Max dose 35mg/kg/day
  - Toxicity-HOLD DRUG
    - ANC <1000
    - Platelet count < 80

Abdominal pain

- Differential Diagnosis
  - Constipation
  - Anxiety
  - Splenic sequestration
  - Gallstones
  - Vaso-occlusive crisis
Abdominal Pain

- **Diagnostic**
  - Palpate for enlarged liver, spleen, constipation
  - Measure spleen or liver and mark
  - Abdominal X-Ray
    - Detect constipation, possibly other findings
  - Ultrasound
    - Detects gallstones
    - More accurate measure of spleen
  - Labs
    - CBC
    - T&H
    - Retic count
    - LFT’s

- **Abdominal X‐Ray**
  - Detect constipation, possibly other findings

- **Ultrasound**
  - More accurate measure of spleen

- **Labs**
  - CBC
  - T&H
  - Retic count
  - LFT’s

- **Diagnostic**
  - Sudden enlarged spleen
  - Hemoglobin at least 2g/dl below baseline
  - Extremely low H & H, low platelets
  - Elevated reticulocyte count and nucleated RBC
  - Infants: severe anemia/hypovolemic shock

- **Treatment**
  - IV Fluids/hypovolemia
  - PRBC transfusion
    - Less blood is given (5cc/kg) (spleen will release)
    - Avoid transfusing to a hemoglobin over 8 g/dl
  - Should see increase in platelet count and retic after transfusion
  - Chronic transfusion (monthly) while has spleen
  - Removal of spleen at later date
  - Pneumococcal 23-valent and meningococcal vaccines 6 weeks before splenectomy
  - Lifelong Penicillin and strict fever precautions after splenectomy

Leg pain/Arm pain

- **Differential diagnosis**
  - AVascular necrosis
  - Humeral head
  - Femoral head
  - Osteomyelitis
  - Vaso-occlusive crisis

Leg/Arm pain

- **Diagnostic**
  - X-ray
  - MRI

- **Labs**
  - CBC
  - Retic count
  - Blood culture
  - CRP

Avascular Necrosis

- **Evaluation**
  - Assess pain
  - Assess joint and mobility
  - MRI

- **Treatment**
  - Pain management
    - NSAIDs
  - Non-weight bearing-crutches
  - Corticosteroid injections-controversial
  - Orthopedic consult
    - Surgery- limited to severe cases and fully grown
Osteomyelitis

- Osteomyelitis
  - Suspect with fever, localized or multifocal bone tenderness especially with erythema and swelling
  - 6 Weeks IV antibiotics
  - Follow blood cultures for ID and sensitivity

Chest Pain

- Differential diagnosis
  - GERD
  - Anxiety
  - Acute Chest Syndrome
  - Vaso-occlusive crisis

Acute Chest Syndrome

- Diagnostic
  - X-Ray
  - O2 saturation
  - CBC, CRP, Retic count, VBG
- Diagnosis of Acute Chest Syndrome
  - Fever
  - New infiltrate on Chest X-Ray
  - SOB, Hypoxia, Chest pain, tachypnea, cough, rales

Acute Chest Syndrome

- Treatment
  - PRBC transfusion
    - Simple transfusion 10 ml/kg unless hgb above 9 g/dl
  - Exchange transfusion
    - Rapid progression of ACS
      - Oxygen saturation below 90 despite supplemental O2
      - Increasing respiratory distress
      - Progressive pulmonary infiltrates
      - Decline in hemoglobin despite simple transfusion
  - CPT
    - Albuterol q 4
    - Incentive Spirometer
    - Cefepime
    - Zithromax
    - Pain management

Headache

- Differential diagnosis
  - Stroke
  - HTN
  - Vaso-occlusive crisis
  - Aplastic Crisis

Stroke

- Headache, altered level of consciousness, numbness tingling, weakness, slurred speech, coma
- If Stroke suspected
  - Urgent neurology consult
  - Urgent consult sickle cell specialist
  - Urgent CT/MRI/MRA
  - Type and cross ASAP
- Treatment
  - Immediate
    - Exchange Transfusion
  - Long term
    - Hydroxyurea
    - Monthly transfusions
    - Compliance with annual or bi-annual transcranial doppler
    - Frequent Ferritin levels
    - X-Jade or Jade new for Ferritins over 1000
Hands/Feet

- Dactylitis
  - Edema and pain to hands and feet
  - Infants, toddlers, preschoolers
    - Can see in as old as 7 yr
  - Treatment
    - Warm compress
    - NSAID’s
    - Fluids
    - Pain management

Penile pain

- Differential Diagnosis
  - Trauma
  - Urinary tract infection
  - Foreskin infection
  - Urethral stone
  - Yeast infection
  - Priapism

Priapism

- Painful erection of penis > 4hrs
  - Stuttering priapism < 4hrs
  - Affects up to 35% of males with sickle cell disease
  - Prompt management can minimize need for invasive treatment
  - Delayed diagnosis and therapy may result in impotence

  - Urologic Emergency

    - Management
      - Surgical intervention immediate
        - Aspiration and irrigation of corpus cavernosum
      - If above not successful
        - Exchange transfusion
      - Fluids
        - Pseudophedrine 30 mg po (30-60 mg per day)
      - Urine
      - Warm shower
      - Analgesia

Fever

- Infants
  - As all other infants
  - Increased risk for infection

  - All ages

    - Diagnostic
      - CBC with diff
      - CRP
      - CP8
        - Type and cross
        - Blood cultures
      - Peripheral
      - Urine culture
      - Chest xray
      - Sputum culture

    - Treatment
      - Admit and start on IV antibiotics follow blood cultures for 48-72 hours
      - Antibiotics
        - Rocephin outpatient (rare)
        - Older healthy child
        - Spleen intact
        - Parents compliant and reliable
        - Local
        - Does not appear overly ill
        - Suspected viral
        - Cefotaxime (Cefotaxime shortage)
        - Follow ID and sensitivity
        - Add Clindamycin or Vancomycin

    - Fever precautions
      - Report to ER or urgent care for all fevers
      - Maintain up to date immunizations
        - Influenza
        - Pneumococcal
        - Meningococcal
      - Prophylactic penicillin
        - All patients under 5 years of age
          - 0-3 years
            - Ose 125 mg po bid
          - 3-5 years
            - Ose 250 mg po bid
        - Indefinitely if Spleen removed

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Hepatobiliary Complications

- Acute Intrahepatic Cholestasis
  - Can be fatal if not recognized and treated promptly
  - Presentation
    - Sudden onset RUQ pain
    - Increasing jaundice
    - Progressively enlarging and tender liver
    - Light colored stools
    - Evidence of hyperbilirubinemia
      - Total serum bilirubin >50 mg/dl
    - Possible thrombocytopenia, hypalbuminemia, elevated alkaline phosphatase, and coagulation abnormalities
  - Management
    - Simple or exchange transfusion
    - Hydration
    - Rest
    - Close observation
    - Immediate consult pediatric hematology/sickle cell disease expert

Differential diagnosis
- Splenic sequestration
- Severe Anemia
- Aplastic Crisis
- Hyperhemolytic crisis
- Infection

Acute Anemia

- Evaluate
  - CBC and reticulocyte count (repeat daily if hospitalized)
- Defined
  - Hemoglobin 2.0g/dl below baseline
  - Or below 6g/dl if baseline unknown
- Have seen baselines hgb as low as 5gm/dl in SS
- Evaluate for etiology requiring urgent intervention
  - Decreased production
    - Low reticulocyte count
    - Aplastic crisis
    - Sequestration
  - Increased destruction
    - Vaso-occlusive crisis
    - Acute chest syndrome
  - Accelerated hemolysis
    - Hyperhemolytic crisis
    - Delayed Hemolytic Transfusion Reaction
  - Blood loss
  - Slow progressive hemoglobin reduction
- Consider renal failure in the older child

Hyperhemolytic Crisis

- Accelerated rate of red blood cell destruction
  - Anemia
  - Jaundice
  - Reticulocytosis

Aplastic Crisis
Aplastic Crisis

- Hemoglobin (3-6 gm/dl)
- Reticulocyte count drastically reduced – zero
- Multiple people in household with sickle cell disease can develop
- Usually caused by prior illness (respir infection)
  - Parvo virus B19
- Re-occurrence very unlikely
- Temporary factory “shut down”- bone marrow

Management: hospital
  - Confirm with CBC and reticulocyte count
  - Febrile work up if fever
  - Blood transfusion
  - Oxygen
  - Rest and recovery

Hepatobiliary complications

- Acute Cholecystitis
  - Not common in SC
  - Can occur with or without presence of gallstones
  - Presentation
    - Colicky pain RUQ with abdominal tenderness on exam
    - Fever
    - Leukocytosis
    - Nausea and Vomiting
  - Nonvisualisation of gallbladder by 60 min after cholecintigraphy
  - Treatment
    - Antibiotics
    - Surgical consult

Hepatobiliary Complications

- Cholelithiasis - presence of gallstones in common bile duct
  - Symptoms
    - Dull pain in RUQ
    - Tender hepatomegaly
    - Rapidly increasing jaundice
  - Management
    - Surgical consult

Acute Ocular Complications

Etiology: Trauma, Infection, Vaso-Occlusive episodes, Proliferative Sickle Retinopathy

- Hyphema
  - Blood in anterior chamber (blunt trauma)
  - Hemorrhage covering lower part of iris
  - Visual abnormalities, floaters, flashes, light sensitivity, blurry vision
- Central Retinal Artery Occlusion
  - Associated with sickle cell disease and moyamoya syndrome, or ACS
  - Causes infarction of inner retina
- Orbital infarction
  - Causes infarction of orbital bones
  - Presentation - proffusion of the eye, pain, eyelid or orbital edema, decreased visual acuity, decreased extraocular motility
- Orbital apex syndrome
  - Optic neuropathy and decreased extraocular motility

Acute Renal Failure

- Defined
  - Rapid reduction in renal function
  - Rise in serum creatinine
  - Reduction in glomerular filtration
  - With or without decline in urine output
- Symptoms
  - Weakness
  - Nausea
  - Fever

- Causes
  - Post nephrotic dehydration
  - Post renal obstruction
  - Intrinsic renal disease glomerular injury
  - Resulting from VOD especially ACS or acute multi system organ failure
  - History of chronic kidney disease and exposure to nephrotoxic medications

Acute Ocular Complications

- Management
  - Prompt examination for hyphema with eye trauma
    - Immediate ophthalmology consult
  - Prompt referral for dilated eye exam
    - Protrusion of eye, changes in visual acuity, flashes or floaters, unilateral bilateral loss of vision
    - Assess for visual acuity, intraocular pressure and peripheral retina
  - Manage acute ocular complications immediately with ophthalmology and hematology (sickle cell specialist)
**Acute Renal Failure**

- Sickle Cell Disease
  - Inability to fully concentrate urine
  - More susceptible to pre-renal azotemia
  - Increased renal tubular secretion
    - Serum creatinine rises only after 30 ml/min or less
    - Values of GFR may be low
- Management
  - Rise in serum creatinine of > 0.3 mg/dl
  - Monitor renal function daily
    - Serum creatine
    - I's and O's
  - Avoid nephrotoxic drugs and imaging agents
  - Evaluate for etiology and consult with nephrologist
- Treatment
  - Exchange transfusion when associated with acute MSOF/Vaso-Occlusion
  - Hemodialysis for other causes

**Discharge**

- If possible coordinate with specialist or PCP (whomever manages sickle cell) with regards to narcotic prescriptions
- Encourage compliance with vaccines
  - Influenza-annual
  - Meningsoccal
  - Pnuemococal
    - Complete series of 13 valent conjugate vaccine-after birth
    - 23 valent pneumococcal polysaccharide vaccine age 2 yr & 5 yrs
- Screenings

**Screening tests**

- Proteinuria
  - Age 10yr annual
- Ophthalmology exam (dilated)
  - Age 10yr (q1-2yr) to evaluate for retinopathy
- Trans Cranial Doppler (TCD)
  - Annual age 2yr-16yr
- PFT's for symptomatic children

**References**

- Up to date. Vasoocclusive pain management in sickle cell disease.  
- NHLBI. Evidence based mgmt of sickle cell disease.  