Evidence-Based Management of Uncomplicated Sickle Cell Disease Vaso-Occlusive Crisis

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Sickle Cell Case Study
25 year old female, history of SCD and AVN bilateral hips. Presents to the ED reporting 10/10 dull, aching pain in back and bilateral hips. Height 5'5" weight 68kg.
• Labs – WBC 12,000, hgb 7.3, reticulocyte count 383, scr 0.3, BP 112/73
• Home pain regimen - morphine ER 90 mg BID, morphine IR 15 mg 1 tab q6h prn (using ATC for the last 2 days)

Objectives
• Define sickle cell disease.
• Explore the prevalence of sickle cell in the U.S.
• Describe the pathology of sickle cell disease.
• Examine the clinical presentation and complications of sickle cell disease and vaso-occlusive crisis (VOC).
• List evidence based treatment strategies for VOC pain management.
• Discuss an evidence-based treatment algorithm and order set for VOC.
Sickle Cell Definition

Definition
Sickle cell disease is a group of inherited red blood cell disorders with an abnormality in the oxygen carrying protein of the hemoglobin.

Autosomal recessive pattern.

(50%) Sickle Cell Trait
(25%) Sickle Cell Disease
(25%) Normal

Definition
Sickle cell trait (HbSA)
Carrier, no expression of disease.

4 common types of sickle cell (HbS) disorders with sickle cell anemia being the most common and most severe.

- HbSS, sickle cell anemia, (normocytic, hemolytic)
- HbSC, (normocytic, hemolytic)
- HbS Beta 0-thalassemia, (microcytic, hemolytic)
- HbS Beta +-thalassemia, (microcytic, hemolytic)

Rare versions HbSD, HbSE, HbSO, severity varies.
Sickle Cell Diagnosis

Diagnosis

Prenatal screening
- Chorionic villus sampling, amniocentesis.

Newborn screening
- Blood smear, red cell morphology.
- Red cell variation
  - Size (anisocytosis), shape (poikilocytosis), color of red cells, sickle shape (Elliotties)
- Howell–Jolly Bodies (small pieces of nuclear material in RBC seen after hemolysis).

SICKLEDEX®
- Solubility test (>6 months old).
  - 10% sickled cells present test is positive.

Hemoglobin electrophoresis
Confirmatory
Differentiates the type of hemoglobin disorder.

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>Sickle cell disease</th>
<th>Sickle cell trait</th>
</tr>
</thead>
<tbody>
<tr>
<td>HbS</td>
<td>0%</td>
<td>80-100%</td>
<td>20-40%</td>
</tr>
<tr>
<td>HbA1</td>
<td>95-98%</td>
<td>0%</td>
<td>60-80%</td>
</tr>
<tr>
<td>HbA2</td>
<td>2-3%</td>
<td>2-3%</td>
<td>2-3%</td>
</tr>
<tr>
<td>HbF</td>
<td>0.8-2%</td>
<td>2%</td>
<td>2%</td>
</tr>
</tbody>
</table>
Sickle Cell Prevalence

Prevalence

Ancestry

- Africa, Middle East, Mediterranean & South Asia.

Sickle Cell Trait

- 1 in 13 African Americans.

Sickle Cell Disease

- 100,000 Americans affected.
- 1 in 365 African Americans births.
- 1 out of every 16,300 Hispanic American births.

Registry and Surveillance System for Hemoglobinopathies (RuSH)
Prevalence

CDC & National Institutes of Health sponsored projects to improve care for the population and to improve data collection.

(RuSH) Registry and Surveillance System for Hemoglobinopathies • CA, FLA, GA, Michigan, NY, NC & Pennsylvania.

(PHRESH) Public Health Research, Epidemiology, and Surveillance for Hemoglobinopathies. CA, GA & Mississippi.

Sickle Cell Data Collection Program, (SCDC) • CA & GA, study on long term trends in diagnosis, treatment and access for SCD.

HP 2020, Sickle cell

Prevalence

Hospital statistics • 230,000 ED visits for pain crisis per year. • Acute care use is estimated at 1.5 billion annual. • A higher risk of death with more than 3 hospitalizations in a year.

Kennestone Regional Medical Center July 2015 – Feb 2016 • 34 Patients admitted with sickle cell crisis. • 17 Multiple admissions. • 12 Readmissions less than 30 days. • 5 Readmissions less than 1 week. • Average 4 patients per month without readmits. • Average LOS=5, highest LOS=23 & lowest LOS=1.

Pathology
Low oxygenation states cause cells to clump together and block blood flow in the capillaries causing pain due to lack of oxygen to the tissues. The cells become irreversibly sickled and have a short life span of ~10-20 days. Anemia occurs as bone marrow production cannot keep up with the breakdown. Sickling factors:
- Hypoxia, high altitude, flying in non-pressurized planes, dehydration, acidosis, Infections, stress.

Clinical Presentation
Clinical Presentation

Anemia: chronic, hemolytic, shortness of breath, tired, dizzy, & pale.
Spleen: function is weakened or destroyed early in life.
Eyes: retina damage common.
Heart disease: enlarged heart, pulmonary hypertension, shortness of breath and fatigue.
Kidneys: trouble concentrating urine, hematuria, infarcts.
Liver & Gall bladder: jaundice, intrahepatic cholestasis, gall stones common.

Leg ulcers: chronic, painful.
Joints: avascular or aseptic necrosis. Bones in hips, knees, ankles, & shoulders affected.
Priapism: prolonged painful erections.
Acute chest syndrome: serious, acute, 2nd most common reason for hospitalization & most common cause of death. Symptoms, SOB, chest pain, fever, cough & tachypnea.
Stroke: silent brain injury versus clinical stroke. One of the most common devastating complications.

Psychosocial
Life long illness with many interactions with health care providers.
Poor self image, negative thoughts and feelings about the condition, stigmatization, cognitive impairments, fears, anxieties, anger, & depression.
Potential for substance abuse and addiction/alcohol abuse, but no more than any other chronic disease.
Concerns regarding, tolerance, dependence, and addiction add complexity to patient care.
Patients report fear of not being believed about their pain, being labeled a "drug seeker," or receiving inadequate pain control when presenting with crisis and this creates more stress/anxiety for the patient.
Psychosocial

High tolerance and physical dependence from long term opioid use combined with the provider’s fear of over-sedation can lead to undertreatment, which can create what’s called a pseudo-addiction.

Tolerance and dependence do not equal addiction.

Sickle cell disease has the same rate of addition as any chronic disease.

Clinical Presentation

Psychosocial

Studies show

Patients were 25 times more likely to report clinicians did a good job with pain management when they perceived the ED physician and site treated them with trust and respect.

85% of physician’s reported they followed opioid retreatment guidelines. High volume providers & those with negative attitudes were less likely to retreat in 30 minutes.

Multidisciplinary pain team: medical, nursing, social services, pharmacology, & psychology.

Vaso-occlusive or pain crisis

Pain resulting from tissue ischemia due to vaso-occlusion. Most common clinical picture in adults. Episode may last for hours, days, or weeks.

- Complex pain, acute on chronic, treat as acute.
  - Nociocceptive hallmark, neuropathic or mixed.
- Pain can occur anywhere and in several places at same time.
  - Most common: Lower back, legs, arms, abdomen, & chest.
  - Sharp, stabbing, throbbing, intense.
- Pain varies from person to person, episode to episode.
- Pain management plans for home and hospital treatment helpful.
Clinical Presentation
Emergent or urgent care
Need to rule out complications.

- Fever greater than 101° F, shortness of breath.
- Chest pain
- Abdominal swelling
- Severe headache, sudden loss of feeling, weakness, or movement.
- Seizure
- Sudden vision problem.
- Painful erection lasting longer than 4 hrs.
- Acute pain anywhere not controlled by home medications.

Clinical Presentation
Diagnostic tests
- CBC with retic count
- BMP
- Liver function test
- Bun, creatinine & serum electrolytes
- Radiology
- Echo & heart cath if cardiac symptoms

Adults annual routine labs: retic, % Hb, renal function, hepatobiliary function, pulmonary function

Evidence Based Treatment
Evidence Based Treatment

Treat the symptoms
- Oxygen therapy
- Pain relieving medications
- Antibiotics
- IV fluids
- Hydroxyurea (HU), FDA approved in 1998 in adults with sickle cell anemia (SCA). Induces formation of HbF.
- Blood transfusions, used for severe cases, iron overload possible with probable heart, & liver failure.
- Erythrocytapheresis, removes HbS and replaces with packed cells.

Cure:
- Hematopoietic stem cell transplantation or allogenic bone marrow transplant.

Evidence Based Treatment

Treat the symptoms
- Medical marijuana
  - California study on Inhaled (4.7% THC/5.1% CBD) vs placebo. ClinicalTrials.gov Identifier: NCT01771731.
- New research:
  - St. Jude Children’s Research Hospital, Found a way to Increase fetal hemoglobin with the use of the CRISPR gene editing to remove a section of DNA that stimulates “gamma-to-beta” switching.

PAIN THERAPY: OPIOID ADVERSE EFFECTS
Constipation
• Develops due to the actions of opioids on receptors throughout the GI tract
  – Decrease in bowel motility and peristalsis
  – Increased anal sphincter tone
• The best prevention is a scheduled laxative regimen
• First line laxatives have stimulant activity
  – Senna (Senokot) tablets
  – Bisacodyl (Dulcolax) tablets
  – Senna/Docusate (Peri-Colace, SennaS, Senna-Plus)

Nausea and Vomiting
• Develops due to the actions of opioids in the chemo trigger zone (CTZ)
• Phenothiazines
  – Compazine 5 – 10 mg PO TID-QID, 25 mg PR BID; Promethazine 12.5 – 25 mg PO/PR/IM Q4-6H PRN
• Serotonin receptor antagonist
  – Ondansetron 4 – 8 mg PO/IV Q8H PRN
• Dopamine antagonist
  – Metoclopramide 5 – 10 mg PO/IV Q6H PRN

Pruritus
• Mechanism is cutaneous mast cell and basophil activation which leads to histamine release
• Antihistamines
  – Diphenhydramine 25 – 50 mg PO Q6H PRN
  – Hydroxyzine 25 mg PO Q6H PRN
• Opioid receptor mixed agonist-antagonist
  – Nalbuphine 2.5 – 5 mg IV
Sedation

- Patient factors that increase risk
  - Sleep disordered breathing, i.e. OSA
  - Obesity
  - Chronic lung disease
  - Opioid naïve
  - PCA demand + continuous rate infusion
  - Advanced age
  - Renal dysfunction (dialysis or CrCl < 30 ml/min)

Pasero Opioid-induced Sedation Scale (POSS)

Pasero C. Assessment of sedation during opioid administration for pain management. JPharmacut Nurs. 2009;24(3):186-90

PAIN THERAPY: OPIOID PHARMACOLOGY
Pain Pathways

Opioid Mechanism of Action

- Interact with opioid receptors (mu, kappa, delta)
- mu receptor is the primary site of action
- Methadone
  - Blocks NMDA receptor
  - Enhances serotonin and norepinephrine activity
- Tramadol (Ultram™, Ultracet™)
  - Enhances serotonin and norepinephrine activity
- Tapentadol (Nucynta™, Nucynta ER™)
  - Enhances norepinephrine activity

Opioid Chemical Classes

- Phenanthrenes
  - Codeine (Tylenol #3)
  - Morphine (MS Contin, Roxanol)
  - Hydrocodone (Vicodin, Lortab, Norco)
  - Hydromorphone (Dilaudid)
  - Oxycodone (OxyCONTIN, Percocet)
  - Oxymorphone (Opana)
  - Buprenorphine (Suboxone, Butrans)
- Diphenylheptanes
  - Methadone
  - Propoxyphene (removed from the US market)
- Phenylpiperidines
  - Fentanyl (Duragesic)
  - Meperidine (Demerol)
- Benzomorphans
  - Pentazocine (Talwin)
- Miscellaneous
  - Tapentadol (Nucynta)
  - Tramadol (Ultram)

True Opioid Allergic Reactions

- Mediated by immunoglobulin E (IgE)
- Symptoms - hives, maculopapular rash, erythema multiforme, pustular rash, severe hypotension, bronchospasm, angioedema, "anaphylaxis-type reactions"
- An opioid from a different opioid class must be used if opioid therapy is necessary

Pseudoallergic Opioid Reactions

- Non-IgE mediated, mechanism is cutaneous mast cell and basophil activation which leads to histamine release
- Symptoms - flushing, sneezing, sweating, exacerbation of asthma, low blood pressure, nausea, vomiting, constipation or somnolence, pruritus and rash
- These adverse effects DO NOT preclude use of morphine.
Opioid Agonists

**Phenanthrenes**
- Codeine
- Tylenol #3™
- Morphine
- MS Contin™, Roxanol™
- Hydrocodone
- Vicodin™, Lortab™, Norco™
- Hydromorphone
- Dilaudid™
- Oxycodone
- OxyCONTIN™, Percocet™
- Oxymorphone
- Opana™

**Diphenylheptanes**
- Methadone
- Propoxyphene
- Darvon™, Darvocet™ (removed from the US market)

**Phenylpiperidines**
- Fentanyl
- Sublimaze™, Duragesic™
- Meperidine
- Demerol™

*Not recommended for pain*

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Opioid Partial & Mixed Agonists

**Partial Agonists**
- Buprenorphine (Butrans™)

**Phenanthrenes**
- Buprenorphine + Naloxone (Suboxone™)

**Mixed Agonist-Antagonists**
- Nalbuphine (Nubain™)

**Benzmorphans**
- Pentazocine+/-Naloxone (Talwin™, Talwin-Nx™)

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Opioid Pharmacokinetics

**Absorption**
- Hydrophillic opioids absorbed in GI tract
  - Morphine, hydrocodone, hydroMORPHONE, oxycodone
- Lipophillic opioids absorbed in gi tract, sublingual, transbucal, transdermal
  - Fentanyl, methadone

**Distribution**
- Hydrophillic opioids remain in blood
- Lipophillic opioids may distribute into fat tissues
  - Methadone extensively distributed into the tissues
Methadone

Opioid Pharmacokinetics

- Metabolism via the liver
  - Active drug metabolites: provide analgesia, may cause negative side effects
  - Inactive drug metabolites: no clinical effects
  - CYP 450 inhibitors/inducers pose risk for drug interactions
    - Inhibitors: slow down metabolism
    - Inducers: speed up metabolism
- Elimination via the kidneys
  - Decreased kidney elimination of active drug metabolites may lead to toxicity

Opioids and Renal Dysfunction

- Majority of opioid analgesics metabolize to active metabolites that are eliminated via the kidneys
  - Exceptions: methadone and fentanyl
- Renal dysfunction leads to accumulation of both the parent compound and active/inactive metabolites
- Prolonged exposure to active metabolites greatly increases risk for respiratory depression, hypotension, or CNS toxicity
- These ADEs ARE PREVENTABLE
### Opioid Metabolism Comparison

<table>
<thead>
<tr>
<th>Drug</th>
<th>Active Metabolite</th>
<th>Inactive Metabolite</th>
<th>CYP 450 Metabolism</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hydrocodone</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Use with caution in moderate to severe kidney impairment</td>
</tr>
<tr>
<td>Morphine</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Not recommended in severe kidney impairment (CKD Stage 4,5 or CrCl &lt; 30 ml/min)</td>
</tr>
<tr>
<td>Oxycodone</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Safer to kidney impairment. Active metabolite has weak clinical effects</td>
</tr>
<tr>
<td>Hydromorphone</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Use with caution in moderate to severe kidney impairment</td>
</tr>
<tr>
<td>Fentanyl</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Safer to kidney impairment. Monitor for CYP450 drug interactions</td>
</tr>
<tr>
<td>Methadone</td>
<td>x</td>
<td>x</td>
<td></td>
<td>Caution in severe liver impairment. Potential for serious CYP 450 drug interactions leading to respiratory depression or heart arrhythmias</td>
</tr>
</tbody>
</table>

Smith H. Opioid Metabolism. Mayo Clinic Proceedings. 2009;84(7):613-624

### Opioid Pharmacodynamic Comparison

<table>
<thead>
<tr>
<th>Drug</th>
<th>Elimination Half-life (hrs)</th>
<th>Duration of Action (hrs)</th>
<th>Dosing Intervals (hrs)</th>
<th>Peak (min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxycodone IR (PO)</td>
<td>2-3</td>
<td>3-6</td>
<td>2-6</td>
<td>60 – 90</td>
</tr>
<tr>
<td>Oxycodone CR (PO)</td>
<td>2-3</td>
<td>3-6</td>
<td>2-6</td>
<td>90 – 180</td>
</tr>
<tr>
<td>Morphine (IV)</td>
<td>2-3</td>
<td>3-4</td>
<td>2-4</td>
<td>15 – 30</td>
</tr>
<tr>
<td>Morphine IR (PO)</td>
<td>2-3</td>
<td>3-4</td>
<td>2-6</td>
<td>90 – 180</td>
</tr>
<tr>
<td>Morphine CR (PO)</td>
<td>2-3</td>
<td>3-6</td>
<td>2-6</td>
<td>90 – 180</td>
</tr>
<tr>
<td>Hydromorphone IR (PO)</td>
<td>2-3</td>
<td>3-4</td>
<td>2-6</td>
<td>10 – 20</td>
</tr>
<tr>
<td>Hydromorphone IR (IV)</td>
<td>2-4</td>
<td>4-8</td>
<td>2-4</td>
<td>30 – 90</td>
</tr>
<tr>
<td>Fentanyl (IV)</td>
<td>2-4</td>
<td>17 (after removal)</td>
<td>2-3</td>
<td>15 – 30</td>
</tr>
<tr>
<td>Fentanyl (TD)</td>
<td>3-4</td>
<td>17 (after removal)</td>
<td>2-3</td>
<td>24</td>
</tr>
<tr>
<td>Methadone (PO)</td>
<td>12-150 hrs</td>
<td>3-6, initially, and 6–12 (or longer) with repeated administration</td>
<td>4 – 6 (initial), 6 – 24 (repeated administration)</td>
<td>150</td>
</tr>
</tbody>
</table>

IR = Immediate release  
CR = Controlled release  
PO = Oral  
IV = Intravenous  
TD = Transdermal

Adapted from UpToDate® 2015: Selected opioid analgesics for pain and equianalgesic doses  
http://www.uptodate.com/contents/image?imageKey=ONC/58864&source=graphics_search&rank=0&search=opioids

Pasero C and McCaffery M. “Pain Assessment and Pharmacologic Management” Chapter 16: Initiating Opioid Therapy, Section IV, Opioid Analgesics. Table 16-1: Equianalgesic Dose Chart

IR = Immediate release  
CR = Controlled release  
PO = Oral  
IV = Intravenous  
TD = Transdermal
Morphine

- Initiate loading doses every 15-30 min until pain is decreased by 30-50%
- Followed by scheduled (ATC) maintenance IV opioid regimen
  - IV PCA or IV bolus doses Q3H ATC
- Continue long-acting oral opioid
  - IV PCA opioid continuous infusion may not be required if long-acting oral is reordered
Calculating Loading Doses

Weight based dosing (opioid naïve and/or < 50kg)

• Calculate the initial IV morphine loading dose at 0.1-0.15mg/kg (max 10mg)

Calculating Loading Doses

Individualized dosing (opioid tolerant)

• Find the morphine equivalent daily dose (MEDD) - convert the current 24 hour ORAL opioid dose to IV morphine
• Reduce by 30% if converting between opioids (incomplete cross-sensitivity)
• Order 10% of the calculated equianalgesic dose as the loading dose
• Morphine IV usual dose 5-10 mg

Equianalgesic Chart

<table>
<thead>
<tr>
<th>Medication</th>
<th>Approximate equianalgesic dose¹</th>
<th>Oral</th>
<th>Parenteral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short-acting opioid agonists</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Morphine (MESI)</td>
<td>50 mg</td>
<td>10 mg</td>
<td></td>
</tr>
<tr>
<td>Codeine²</td>
<td>200 mg</td>
<td>100 mg</td>
<td></td>
</tr>
<tr>
<td>Hydrocodone² (Oxycodone)</td>
<td>7.5 mg</td>
<td>1.5 mg</td>
<td></td>
</tr>
<tr>
<td>Meperidine² (Demerol) NR</td>
<td>300 mg</td>
<td>100 mg</td>
<td></td>
</tr>
<tr>
<td>Oxymorphone² (Normorphine)</td>
<td>N/A</td>
<td>1 mg</td>
<td></td>
</tr>
<tr>
<td>Oxycodeone (Oxycodeone, OXYIR)</td>
<td>30 mg</td>
<td>N/A</td>
<td></td>
</tr>
</tbody>
</table>

2 American Pain Society Sickle Cell Guidelines
**Initial PCA Demand Doses**

<table>
<thead>
<tr>
<th>Less than 50kg</th>
<th>Usual start dose after loading dose</th>
<th>Usual dose range</th>
<th>Lockout interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine IV</td>
<td>0.02 mg/kg/dose</td>
<td>0.01-0.03 mg/kg/dose</td>
<td>6-10 minutes</td>
</tr>
<tr>
<td>Dilaudid IV</td>
<td>0.003-0.004 mg/kg/dose</td>
<td>0.003-0.005 mg/kg/dose</td>
<td>6-10 minutes</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Greater than 50kg</th>
<th>Usual start dose after loading dose</th>
<th>Usual dose range</th>
<th>Lockout interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morphine IV</td>
<td>1.0 mg</td>
<td>0.5-2.5 mg</td>
<td>6-10 minutes</td>
</tr>
<tr>
<td>Dilaudid IV</td>
<td>0.2 mg</td>
<td>0.05-0.4 mg</td>
<td>6-10 minutes</td>
</tr>
</tbody>
</table>

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**Opioid Weaning**

- PCA weaning
  - If continuous rate infusion begin decreasing this first
  - Next decrease the demand dose and/or change the lockout interval
- Resume oral short-acting opioid

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**PAIN THERAPY: NON-OPIOID MODALITIES**
NSAIDs and Acetaminophen

- Schedule around-the-clock
- NSAIDs avoid in AKI (CrCl < 30ml/min)
  - Ketorolac 15-30 mg IV Q6H
  - Ibuprofen 600 mg ORAL Q6H
- Acetaminophen 500-1000 mg ORAL Q6H
  - Avoid if liver enzymes are significantly elevated

Ketamine

- Clinical role in sickle cell pain therapy
  - Second line therapy for acute pain refractory to opioids
- Mechanism of action
  - N-methyl-d-aspartate (NMDA) receptor antagonist
  - Analgesic, sedative and amnestic properties
  - Analgesia occurs at lower doses than anesthetic and psychotomimetic effects

Ketamine

- Dosing
  - 0.2 – 0.5 mg/kg IV bolus dose over 10minutes
- Contraindications
  - head trauma, post intracranial surgery, increased intracranial pressure, intracranial bleeding, intracranial mass, seizure disorder
- Precautions
  - Hyper/hypotension, PTSD, psychosis/schizophrenia, history of stroke or MI
Ketamine

- **Adverse effects**
  - Psychomimetic – hallucinations, dream-like feelings; co-administration of lorazepam of low-dose haloperidol is recommend for prevention

- **Clinical monitoring**
  - Vital signs including respiratory status, pain sedation, adverse effects every 15 minutes x 1 hr, then every 2 hours x 2 hours

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Hydration

- Prevent/reverse dehydration, decrease sodium concentration outside the red cells
- Encourage oral fluids. If unable to drink fluids, provide intravenous hydration at 125ml/hr to avoid over-hydration.
- Hypotonic solutions - D5W or D5 with ¼ or ½ NS.

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Oxygen

- Prevent deoxygenation – primary cause of sickling
- Administer oxygen to all patients with hypoxia (oxygen saturation < 95 % on room air) or dyspnea
Complimentary Therapy

- Adjunctive non-pharmacologic approaches to treat pain
  - Heat therapy
  - Distraction – TV, music, relaxation

ACS Risk Reduction

- Reduce risk of acute chest syndrome
  - Encourage incentive spirometry use while awake
  - Encourage ambulation and activity as soon as possible

ABCs for Managing Sickle Cell Pain

<table>
<thead>
<tr>
<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
<th>E</th>
<th>F</th>
</tr>
</thead>
<tbody>
<tr>
<td>Assessment of the pain (see pain management tool)</td>
<td>Believe the patient’s level of pain</td>
<td>Complications or cause of pain (look for complications)</td>
<td>Drugs and Distraction</td>
<td>Pain Medication (opioids and NSARCH, if no contraindications)</td>
<td>Distraction with music, TV, relaxation techniques</td>
</tr>
<tr>
<td>Environmental, not in quiet area with privacy</td>
<td>Nonpharmacologic (e.g., DS with 0.25 normal saline solution)</td>
<td>Use fixed dosing given on a time schedule: no prn dosing for pain medications</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


### Evidence Based Algorithm

- Identified the need
- Literature review
- Team formed
- Order set & pathway developed
- Approval process
- Electronic record journey
- Education

### EBP Treatment Algorithm & Order Set

**Process**

- Identified the need
- Literature review
- Team formed
- Order set & pathway developed
- Approval process
- Electronic record journey
- Education

**Next steps**

- ED specific order set
- Formalize transition to outpatient
Sickle Cell Case Study

25 year old female, history of SCD and AVN bilateral hips. Presents to the ED reporting 10/10 dull, aching pain in back and bilateral hips. Height 5'5" weight 68kg.

- Labs – WBC 12,000, hgb 7.3, reticulocyte count 383, scr 0.3, BP 112/73
- Home pain regimen - morphine ER 90 mg BID, morphine IR 15 mg 1 tab q6h pm (using ATC for the last 2 days)

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