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

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we believe in life well-lived. WELLSTAR.

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)

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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.

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Sickle Cell Definition

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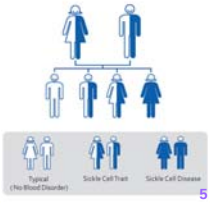
Definition

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



https://www.cdc.gov/ncbddd/sicklecell/traits.html

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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.

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Sickle Cell Diagnosis

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Diagnosis

Prenatal screening

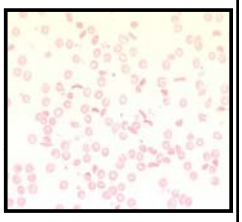
- Chorionic villus sampling, amniocentesis.

Newborn screening

- Blood smear, red cell morphology.
- Red cell variation
  - Size (anisocytosis), shape (poikilocytosis), color of red cells, sickle shape (Elliptocytes).
  - 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.



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Diagnosis

Hemoglobin electrophoresis

Confirmatory

Differentiates the type of hemoglobin disorder.

	Normal	Sickle cell disease	Sickle cell trait
HbS	0%	80-100%	20-40%
HbA1	95-98%	0%	60-80%
HbA2	2-3%	2-3%	2-3%
HbF	0.8-2%	2%	2%

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Sickle Cell Prevalence

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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.

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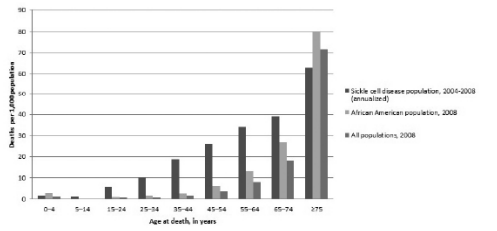
Prevalence

Registry and Surveillance System for Hemoglobinopathies (RuSH)

Figure 1. All-cause mortality rates for the sickle cell disease population identified through population-based surveillance (2004 through 2008), the African American population (2008),* and all populations (2008),* California and Georgia



*Centers for Disease Control and Prevention (US), National Center for Health Statistics. Multiple cause of death 1999–2013 [cited 2015 Dec 15]. Available from: http://wonder.cdc.gov/mcd.html

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Prevalence

CDC & National Institutes of Heath 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

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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 10 patients per month.
- Average 4 patients per month without readmits.
- Average LOS=5, highest LOS=23 & lowest LOS=1.

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Pathology

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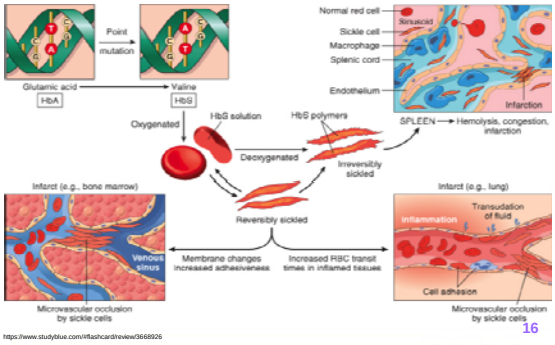
Pathology



https://www.studyblue.com/#flashcard/review/3668926

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Pathology

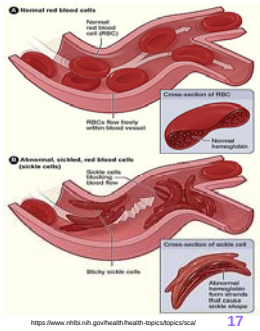
Low oxygenation states cause cells to clump together and block blood flow in the capillaries causing pain due to lack of oxygen to the tissues.

The cells become irreversibly sickled and have a short life span of ~10-20 days.

Anemia occurs as bone marrow production can not keep up with the breakdown.

Sickling factors:

- Hypoxia, high altitude, flying in non-pressurized planes, dehydration, acidosis, Infections, stress.



https://www.nhlbi.nih.gov/health/health-topics/topics/sca/

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Clinical Presentation

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Clinical Presentation

Anemia: chronic, hemolytic, shortness of breath, tired, dizzy, & pale.

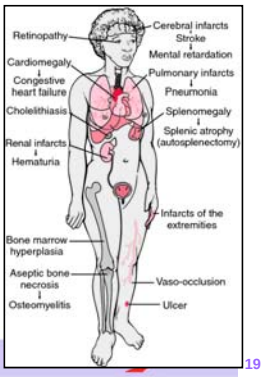
Spleen: function is weakened or destroyed early in life.

Eyes: retina damage common.

Heart disease: enlarged heart, pulmonary hypertension, shortness of breath and fatigue.

Kidneys: trouble concentrating urine, hematuria, infarcts.

Liver & Gall bladder: jaundiced, intrahepatic cholestasis, gall stones common.



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Clinicopathologic findings in sickle cell anemia. The findings are a consequence of infarctions, anemia, hemolysis, and recurrent infection. From Damjanov, 2000. Found on the free dictionary sight

Clinical Presentation

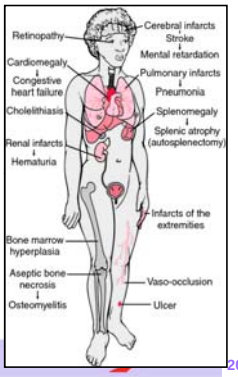
Leg ulcers: chronic, painful.

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.



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Clinicopathologic findings in sickle cell anemia. The findings are a consequence of infarctions, anemia, hemolysis, and recurrent infection. From Damjanov, 2000. Found on the free dictionary sight

Clinical Presentation

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.

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Clinical Presentation

Psychosocial

High tolerance and physical dependence from long term opioid use combined with the provider's fear of over-sedation can lead to under treatment, 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.

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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.

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Clinical Presentation

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.
  - Nocioceptive 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.

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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.

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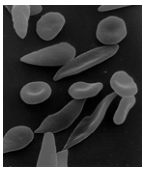
Clinical Presentation

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

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Scanning electron micrograph showing a mixture of red blood cells, some with round normal morphology, some with mild sickling showing elongation and bending By NIDDK - (US government agency) site at http://www.cc.nih.gov/ccc/ccnews/nov99/ . The photo is attributed to Drs. Noguchi, Rodgers, and Schechter of NIDDK., Public Domain, https://commons.wikimedia.org/w/index.php?curid=4630192

Evidence Based Treatment

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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.

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Evidence Based Treatment

Treat the symptoms
- Medical marijuana
  - California study on Inhaled (4.7% THC/5.1% CBD) vs placebo. ClinicalTrials.gov Identifier: NCT01771731.
  - Medical marijuana site, 16 states with laws on use. http://medicalmarijuana.procon.org/view.resource.php?resourceID=000881
- 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.

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PAIN THERAPY: OPIOID ADVERSE EFFECTS

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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)

Goodheart CR, Leavitt SB. Managing Opioid-Induced Constipation in Ambulatory-Care Patients. August 2006. Available online at www.Pain-Topix.com.

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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/IV/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

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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

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Januzzi R. Nalbuphine for treatment of opioid-induced pruritis: a systematic review of literature.ClinJPain.Jan2016;32(1):87-93.

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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)

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Pasero Opioid-induced Sedation Scale (POSS)

Table 1. Pasero Opioid-induced Sedation Scale (POSS) With Interventions*

S = Sleep, easy to arouse
Acceptable; no action necessary; may increase opioid dose if needed
1. Awake and alert
Acceptable; no action necessary; may increase opioid dose if needed
2. Slightly drowsy, easily aroused
Acceptable; no action necessary; may increase opioid dose if needed
3. Frequently drowsy, arousable, drifts off to sleep during conversation
Unacceptable; monitor respiratory status and sedation level closely until sedation level is stable at less than 3 and respiratory status is satisfactory; decrease opioid dose 25% to 50%[1] or notify prescriber[2] or anesthesiologist for orders; consider administering a non-sedating, opioid-sparing nonopioid, such as acetaminophen or an NSAID, if not contraindicated.
4. Somnolent, minimal or no response to verbal or physical stimulation
Unacceptable; stop opioid; consider administering naloxone[3,4]; notify prescriber[2] or anesthesiologist; monitor respiratory status and sedation level closely until sedation level is stable at less than 3 and respiratory status is satisfactory.

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

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PAIN THERAPY: OPIOID PHARMACOLOGY

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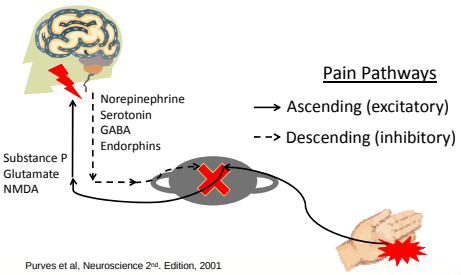
Pain Pathways



Purves et al, Neuroscience 2nd. Edition, 2001

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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

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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)
- Benzmorphans
  - Pentazocine (Talwin)
- Miscellaneous
  - Tapentadol (Nucynta)
  - Tramadol (Ultram)

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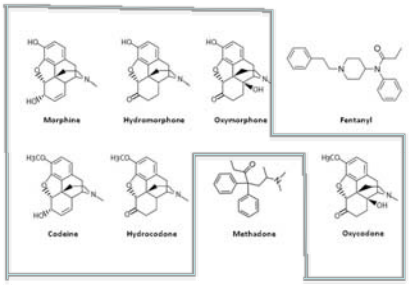
Trescot, et al. Pain Physician, 2008;SI11:S133-S153

Opioid Chemical Structures



http://www.elearn.ascp.org/AngelUploads/Content/LQCL1301/_assoc/CL_13-01.html

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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

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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.**

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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

Phenanthrenes
- Buprenorphine (Butrans™)

Miscellaneous
- Tapentadol (Nucynta™)
- Tramadol (Ultram™)

Mixed Agonist-Antagonists

Pheneanthrenes
- Buprenorphine + Naloxone (Suboxone™)
- Nalbuphine (Nubain™)
- Butrophanol
- Levorphanol

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

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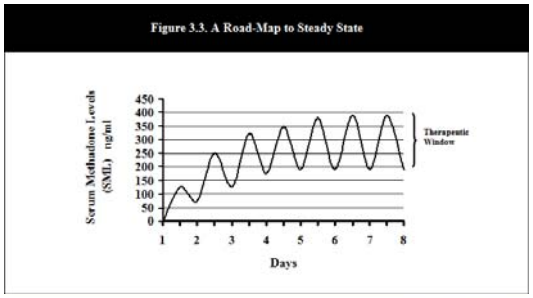
Methadone



http://www.mededta.org/curriculum/curriculum_c03.htm

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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

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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

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Opioid Metabolism Comparison

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

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

Opioid Pharmacodynamic Comparison

Drug	Elimination Half-life (hrs)	Duration of Action (hrs)	Dosing Intervals (hrs)	Peak (min)
Oxycodone IR (PO) Oxycodone CR (PO)	2-3	3-6 8-12	2 - 6 12	60 – 90 90 - 180
Morphine (IV) **Morphine IR (PO)** **Morphine CR (PO)**	**2-3** **2-3**	**3-4** **3-6** **8-12**	**2 – 4** **2 – 6** **8 - 12**	**15 – 30** **60 – 90** **90 - 180**
Hydromorphone (IV) Hydromorphone IR (PO)	2-3 2-3	3-4 3-6	2 – 4 2 – 6	10 – 20 30 - 90
Fentanyl (IV) Fentanyl (TD)	3 – 4 17 (after removal)	2 48-72, up to 12 after removal	2 - 3 72	15 – 30 24
Methadone (PO)	12-150 hrs	3-6, initially, and 8 – 12 (or longer) with repeated administration	4 – 6 (initial) 8 – 24 (repeated administration)	150

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/58964&source=graphics_search&rank=0&searchId=
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

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PAIN THERAPY: MORPHINE DOSING STRATEGIES FOR SICKLE CELL PAIN CRISIS

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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

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U.S. Department of Health and Human Services.National Institutes of Health, National Heart, Lung, Blood Institute. Evidence-based management of sickle cell disease. Expert Panel Report, 2014.

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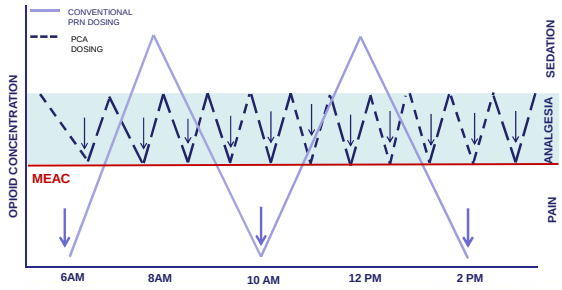
Minimal Effective Analgesic Concentration and the Analgesic Corridor



Anesth Analg 2005;101:S44–S61

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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)

55

American Pain Society.Principles of analgesic use in the treatment of acute pain and cancer pain.6th edition.2008

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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

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American Pain Society.Principles of analgesic use in the treatment of acute pain and cancer pain.6th edition.2008

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Equianalgesic Chart

Table 7. Equianalgesic Doses for Opioid Analgesics in Opioid-Naive Adults and Children ≥50 kg Body Weight[1]

Medication	Approximate equianalgesic dose[1]		
	Oral	≅	Parenteral
Short-acting opioid agonists			
Morphine[2] (MSIR)	30 mg		10 mg
Codeine[3]	200 mg		120 mg
Hydromorphone[2] (Dilaudid)	7.5 mg		1.5 mg
Meperidine[5] (Demerol) N/R	300 mg		100 mg
Oxymorphone[2] (Numorphone)	N/A		1 mg
Oxycodone (Roxicodone, OXY1R)	30 mg		N/A

American Pain Society Sickle Cell Guidelines

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Initial PCA Demand Doses

Less than 50kg	Usual start dose after loading dose	Usual dose range	Lockout Interval:
Morphine IV	0.02 mg/kg/dose	0.01-0.03 mg/kg/dose	6 -10 minutes
Dilaudid IV	0.003-0.004 mg/kg/dose	0.003-0.005 mg/kg/dose	6 -10 minutes

Greater than 50kg	Usual start dose after loading dose	Usual dose range	Lockout Interval:
Morphine IV	1.0 mg	0.5-2.5 mg	6-10 minutes
Dilaudid IV	0.2 mg	0.05-0.4 mg	6-10 minutes

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American Pain Society.Principles of analgesic use in the treatment of acute pain and cancer pain.6th edition.2008

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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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American Pain Society.Principles of analgesic use in the treatment of acute pain and cancer pain.6th edition.2008

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PAIN THERAPY: NON-OPIOID MODALITIES

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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

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Platt A, Eckman J, Beasley J. Treating sickle cell pain: an update from the Georgia comprehensive sickle cell center. JEmergNurs. Aug 2002;28(4):297-303.

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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

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Pasero C, McCaffery M. Pain assessment and pharmacologic management. Chapter 23.pp 674-77.St. Louis.MosbyElsevier.2011.

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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

63

Pasero C, McCaffery M. Pain assessment and pharmacologic management. Chapter 23.pp 674-77.St. Louis.MosbyElsevier.2011.

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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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Pasero C, McCaffery M. Pain assessment and pharmacologic management. Chapter 23.pp 674-77.St. Louis.MosbyElsevier.2011.

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.

65

Platt A, Eckman J, Beasley J. Treating sickle cell pain: an update from the Georgia comprehensive sickle cell center. JEmergNurs. Aug 2002;28(4):297-303.
U.S. Department of Health and Human Services.National Institutes of Health, National Heart, Lung, Blood Institute. Evidence-based management of sickle cell disease. Expert Panel Report, 2014.

Oxygen

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

66

Platt A, Eckman J, Beasley J. Treating sickle cell pain: an update from the Georgia comprehensive sickle cell center. JEmergNurs. Aug 2002;28(4):297-303.
U.S. Department of Health and Human Services.National Institutes of Health, National Heart, Lung, Blood Institute. Evidence-based management of sickle cell disease. Expert Panel Report, 2014.

Complimentary Therapy

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

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Platt A, Eckman J, Beasley J. Treating sickle cell pain: an update from the Georgia comprehensive sickle cell center. JEmergNurs. Aug 2002;28(4):297-303.
U.S. Department of Health and Human Services.National Institutes of Health, National Heart, Lung, Blood Institute. Evidence-based management of sickle cell disease. Expert Panel Report, 2014.

ACS Risk Reduction

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

68

U.S. Department of Health and Human Services.National Institutes of Health, National Heart, Lung, Blood Institute. Evidence-based management of sickle cell disease. Expert Panel Report, 2014.

ABCs for Managing Sickle Cell Pain

TABLE 2
The ABC's for managing sickle cell pain

A —Assessment of the pain (use a pain assessment tool)
B —Believe the patient's level of pain
C—Complications or cause of pain (look for complications)
D—Drugs and distraction
—Pain Medication (opiates and NSAIDs, if no contraindications)
—Distraction with music, TV, relaxation techniques
E —Environment, rest in quiet area with privacy
F —Fluids (Hypotonic-D,W or D5 with 0.25 normal saline solution)
—Use fixed dosing; give on a time schedule; no prn dosing for pain medications

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Platt A, Eckman J, Beasley J. Treating sickle cell pain: an update from the Georgia comprehensive sickle cell center. JEmergNurs. Aug 2002;28(4):297-303.

Evidence Based Algorithm

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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

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EBP Treatment Algorithm & Order Set

Sickle Cell Pain Crisis Order Set

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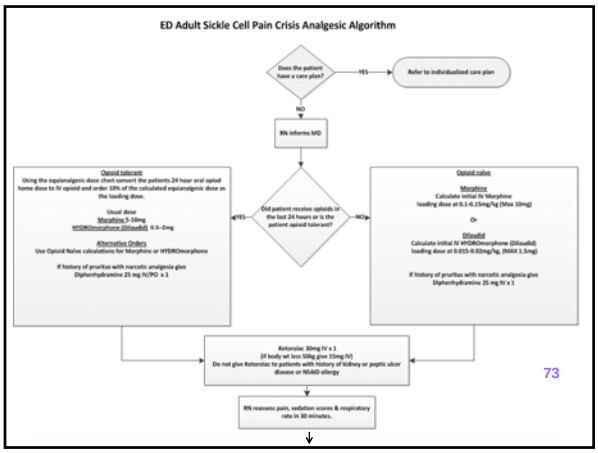

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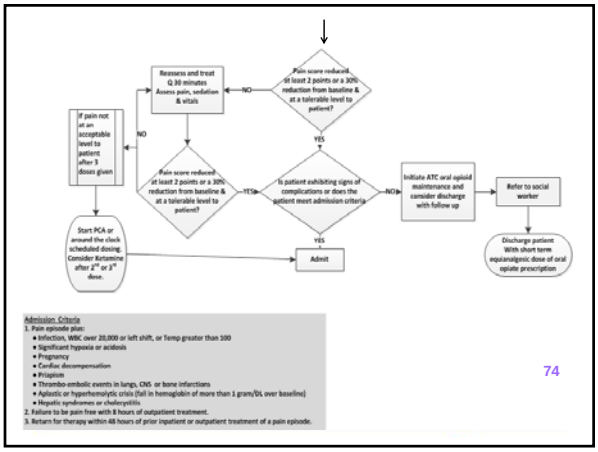

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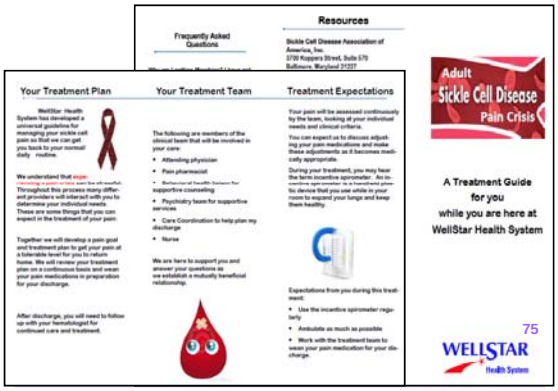
Patient Brochure



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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)

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