AMERICAN COLLEGE OF EMERGENCY PHYSICIANS
SCIENTIFIC ASSEMBLY
1999

Integrated Approach to the Emergency Management of Sickle Cell Anemia Pain Crisis

For the emergency physician, sickle cell disease is most important hemoglobinopathy; patients with this disease are seen almost daily in most emergency departments. Using an evidence-based medicine approach, the most efficient workup of these patients will be discussed, based on presenting complaint. We will dispel some of the common misconceptions in the treatment of sickle cell disorders with regard to oxygen, fluids, and pain medicine. In addition, a multidisciplinary approach to the management of these patients will be presented.

- Understand the various sickling disorders and their implications for complications.
- Learn management of the various crises that occur in patients with sickle cell disease.
- Discuss the admission criteria for presenting complaints, including fever, pain, and anemia.
- Describe a multidisciplinary approach to the management of sickle cell pain and its benefits.

WE-172
Wednesday, October 13, 1999
4:00 PM - 4:55 PM
Room # N242
Las Vegas Convention Center

FACULTY

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Integrated Approach to the Emergency Management of Sickle Cell Anemia Pain Crisis

ACEP Scientific Assembly - Wednesday, October 13, 1999 - (WE172)
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Sickle Cell Pain "Crisis"

Physician Crisis

Multi-Disciplinary Approach to Sickle Cell Patient Management

Todd B. Taylor, MD, FACEP

Clinical Case (Final Exam)

28 YOBM, who recently moved from back East where he had been treated by a pain specialist under a "pain contract".

Upon arriving to Arizona, he immediately applied for AHCCCS (Medicaid) and was assigned to a PCP who confirmed the "pain contract" and has been honoring its protocol.

Clinical Case (Final Exam)

Since arriving two months ago, the patient has been to the ED 10 times, the PCP's office 12 times, and admitted 5 times with "sickle cell crisis". Total hospital days have been 18 days out of the last 60 days and he has received an average of 1.75gm of Demerol per hospital admission (8.75gm total) and 250mg of Demerol per ED\office visit (5.5gm total). Total Demerol in 60 days was 14.25gm or almost 10mg per hour. He has experienced no serious illness or complication during this period of time. He has been maintained on hydroxyurea and claims to be a Jehovah Witness and refuses blood transfusion.

Clinical Case (Final Exam)

The patient has been referred to you for "consultation" and management of this "difficult case".

What are your recommendations?

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143 Page Handbook
by
James R. Eckman, M.D.

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Director, Georgia Sickle Cell Center
Emory Univ. School of Medicine

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Problem Oriented Management of Sickle Syndromes
{graphic}

Georgia Sickle Cell Center

Grady Memorial Hospital
Atlanta, Georgia

- Emergency, Primary, & Tertiary Sickle Cell Care
- Over 1100 patients treated since 1985
- 24 hour / day basis

Sickle Syndromes

- Common Medical Problems
- Complications of Sickle Cell Disease
- Combinations of both

Progress in Sickle Cell Disease

Median Age at Death

- Used to be primarily "pediatric" disease
- 1972
  - 14 years
- 1993
  - 42 years for men
  - 47 years for women

History of Sickle Cell Disease

- 1910
  - The first case of Sickle Cell Anemia
  - "Peculiar Elongated & Sickled Shaped Corpuscles in a Case of Severe Anemia"
- 1927 - Hahn & Gillespie
  - Deoxygenation shown to be primary stimulus for sickling
- 1949 - Linus Pauling
  - Demonstrated the cause by an abnormal hemoglobin
- 1956 - V. M. Ingram
  - Substitution of glutamic acid by valine in the 6th amino acid position of the beta polypeptide chain.

Inherited Sickle Cell Syndromes

- Normal Types of Hemoglobin
  - Hb A ($\alpha_2\beta_2$) - Most common after 6 months of age
  - Hb F ($\alpha_2\gamma_2$) - Fetal hemoglobin
  - Hb A_2 ($\alpha_2\delta_2$)

Inherited Sickle Cell Syndromes

- Homozygous Hemoglobin S
  - Hb S
  - Glutamic Acid substitution by Valine
- Heterozygosity
  - Hb SC (Compound Heterozygosity)
    - One hemoglobin S & one hemoglobin C gene
    - Hb C - Glutamic Acid substitution by Lysine
  - Hb SA
    - One hemoglobin A & one hemoglobin S gene
    - Sickle Cell "Trait" or "Carrier"

Glutamic Acid Substitution by Valine

- Allows the hemoglobin molecule to polymerize
- Hb S forms long tactoids distorting the red blood cell shape & cell membrane
- Increases blood viscosity
- Sludging in end arterioles leads to ischemia & microvascular infarction.

Inherited Sickle Cell Syndromes

Thalassemia

- Beta Thalassemia
  - Beta^0 Thalassemia "Major"
    - Homozygous β-chain thalassemia
    - Absence of normal Hb A
    - Mediterranean populations
  - Beta$^+$ Thalassemia "Minor"
    - Heterozygous β-chain thalassemia
    - Markedly reduced normal Hb A
- Alpha Thalassemia
  - Wide spectrum of disease: Asymptomatic to prenatal death
  - Oriental & black populations

Inherited Sickle Cell Syndromes

Thalassemia - Sickle Cell Combination

- Hb S β^0 thal
  - No production of normal hemoglobin
  - Clinical syndrome similar to sickle cell anemia (Hb SS)
- Hb S β^+ thal
  - Production of 5-20% normal Hb A
  - Clinical syndrome similar to sickle cell "trait" (Hb SA)
  - Clinical symptoms usually much more mild

Sickle Cell Demographics

- Higher frequency in descendents from geographic areas where malaria is endemic.
- Carriers with abnormal Hb or thalassemia show protection against malarial infection.
- Highest frequency populations:

Africans	Arabs
Egyptians	Turks
Greeks	Italians
Iranians	Asiatic Indians

Sickle Cell Demographics

African Americans

- Most common Hb SS group in America
- Carriers
  - 8 % - Hb S
  - 3 % - Hb C
  - 1½ % - Beta Thalassemia
- Incidence live black births
  - Hb SS - 1\625
  - Hb SC - 1\833
  - Hb S β thal - 1\1667

Diagnosis

- Hemoglobin Electrophoresis
- Sickledex
  - Excludes Sickle Cell in those > 6 months without severe anemia or high levels of Hb F
  - Positive if > 10% Hb S
  - Does not differentiate:
    Hb SS, Hb SA, Hb SC, Hb S β thal
  - Does not detect carriers of Hb C or Thalassemias

Clinical Manifestations

- Caused by altered solubility characteristics of genetically abnormal Hgb
  - Changes in RBC deformability & fragility
  - Increase in blood viscosity leading to:
    - Episodic vascular occlusions
    - Ischemic tissue
    - Pain crisis
    - Ultimately end organ failure
  - RBC membrane changes contribute to:
    - Hemoglobin polymerization
    - Adherence of RBC's to vascular endothelium
    - Phagocytosis by macrophage

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Polymerization of Hb S

Factor	Sickling Increased	Sickling Decreased
O_2 Sat	Deoxygenation	Oxygenation
Hb S Conc	High % Hb S High MCHC Dehydration	High % Hb F Low MCHC Hyponatremia
Temp	Fever	Euthermia
Blood pH	Acidosis	Alkalosis

Clinical Manifestations

- Results
  - Chronic Hemolytic Anemia
  - Increased susceptibility to serious infection
  - Wide-spread ischemic damage
  - End organ failure

Early Symptoms

- Newborn screening identifies most
- Jaundice, irritability, colic, failure to thrive, fever, vomiting
- Hepatosplenomegaly, pallor, heart murmurs
- Overwhelming sepsis

Anemia

- Lifelong
- Worsens as Hb F falls in first year of life.
- Average RBC life-span reduced from 120 days to 10-20 days.
- Elevate reticulocyte count
- Proliferation of RBC precursors
- Jaundice (elevated indirect bilirubin)
- Changes in bone structure
- Elevated LDH

Anemia

Admission Criteria

- Unstable Hgb < 6gm%
- Drop from baseline of > 1gm% with:
  - Heart failure
  - Pain crisis
  - GI or GU bleeding
  - Significant orthostatic blood pressure changes
  - Infection

Transfusion Therapy

- Goals:
  - Relieve symptoms
  - Treat & prevent complications
  - Dilute out sickle cells with normal cells
- Should be considered when anemia is severe enough to compromise tissue oxygenation with complications.
  - Pulmonary or cardiac compromise
  - Prevent recurrent CNS events
- Life-Saving
  - Aplastic crisis
  - Sequestration crisis

Transfusion Criteria

- Decision to transfuse is based more on a change from baseline or presence of complication than an absolute level.

Transfusion Criteria

- Definite Indications:
  - Acute neurological event
  - Splenic\hepatic sequestration
  - Severe pneumonia or pulmonary infarction
  - Severe anemia with cardiac decompensation
  - Acute hypoxia
  - Aplastic crisis with anemia
  - Hyperhemolytic crisis with enlarging liver\spleen
  - Ophthalmological surgery

Transfusion Criteria

- Relative Indications:
  - Symptomatic anemia
  - Refractory leg ulcers
  - Refractory priapism
  - Frequent or severe or prolonged pain episode
  - Chronic respiratory insufficiency
  - High dose intravascular contrast studies
  - General anesthesia
  - Pregnancy

Transfusion Criteria

- Usually Not Indicated:
  - Stable chronic anemia
  - Pain episode without prior criteria
  - Minor surgery with local or short general anesthesia
  - Uncomplicated infections
  - Chronic bone disease or organ failure without other indication

Transfusion Therapy

- Admission Criteria:
  - Patients with absolute transfusion requirement
  - Acute exchange transfusion indication
  - Underlying CHF or uncontrolled hypertension
- Outpatient Criteria
  - Stable chronic transfusion program
  - Preparation for elective surgery or contrast studies

Transfusion Therapy Considerations

- Infectious Diseases
  - Hepatitis, HIV, CMV, etc
- Volume Overload
  - Plasma volume usually already expanded with chronic anemia
- Iron Overload
- Alloimmunization
  - Increased due to exposure to antigens missing in abnormal sickle cells
- Whole Blood Viscosity

Transfusion Therapy

- Whole Blood Viscosity in Mixed Sickle & Normal Cells
  - Increased viscosity until normal cells > 50%
- Hct > 30% & Normal Cells < 50%
  - Viscosity rapidly increases
  - Clinical symptoms appear when Hct > 35%
  - Rarely necessary to raise Hct >35%

Transfusion Therapy

- Simple Transfusion: PRBC's
  - Used to raise Hct only up to 30% initially
  - Goal is to increases percentage of Hb A > 50% over time
  - Not intended to increase Hct > 35%
  - Chronic transfusion every 3-4 weeks to maintain Hct > 30%, Hb A > 70%, & ARC < 4%
- Exchange Transfusion
  - Used when the goal is to increase Hct up to 35% rapidly
  - Hb A level of > 50% should also be achieved

Iron & Folate Deficiency

- Iron Deficiency
  - Increasing anemia with decreasing ARC &\or MCV
  - Serum ferritin < 20ng/ml is diagnostic
- Folate Deficiency
  - Increasing anemia with increasing MCV, low ARC, & low serum\RBC folate

Clinical Case

A 3 year old black male with known Hb SS disease presents to the ED carried by his mother. This child is pale, listless, & floppy.

VS: HR-150 RR-28 BP-Unobtainable

What do you do next?

Sequestration Episode (Crisis)

- Occurrence
  - Hb SS - Most common less than 5 years of age
  - Other types - Any age
- Characterized By:
  - Life-threatening anemia
  - Rapid enlargement of the spleen &\or liver
  - Increasing jaundice
  - High reticulocyte count (>100K)
  - Hgb drop of > 1gm% from baseline or absolute Hgb < 5gm%
  - Elevated WBC & platelets
  - Severe pain episode
  - +\- Hypotension & pallor
- Older may have primarily liver sequestration
  - Rapid deterioration of liver function
  - Rhabdomyolysis
  - Renal insufficiency

Sequestration Episode (Crisis)

Treatment

- Rapid exchange transfusion
- Occasional emergent splenectomy
- Tend to be recurrent
- Children < 5 Years
  - Transfuse every 3-4 weeks to maintain Hgb 11gm%, ARC < 4%, & Hb S < 30%
  - Elective splenectomy at age 5
- Children > 5 years
  - Elective splenectomy after first or second episode

Aplastic Episode (Crisis)

- Most frequently during early childhood
- Rapid onset of life-threatening anemia
- Due to decreased or absence of RBC production
- Reticulocyte count is low (<10K), absent or markedly decrease RBC precursors in bone marrow with a drop in Hgb of >1gm% from baseline or Hgb < 5gm%.
- Can be precipitated by parvovirus B19 infection
- Any stress, particularly infection, can cause increased hemolysis and decreased RBC production
- Treatment involves transfusion support and observation.

Infection

- Major cause of death - particularly < 3 years old
- Encapsulated organisms from "autosplenectomy" early in life
  - Streptococcus pneumoniae
  - Salmonella
  - Hemophilus influenza
  - Meningococcus
- Bone & joint infections are more common due to ischemia
- Urinary tract infections

Infection

Treatment

- Routine & Special Immunizations
  - Pneumococcal vaccine
  - Hemophilus B vaccine
- Empiric Antibiotics
  - Significant fever
  - Documented infection

Stroke

- 80% Occur before the age of 12
- 85% Recurrence within 3 years
- May present as:
  - Hemiplegia
  - Seizure
  - TIA
  - Coma
  - Sensory loss
- Treatment
  - Transfusion every 3-4 weeks to maintain Hb S levels < 30%, Hgb 11gm / dl, ARC < 4%

Dactylitis: Hand - Foot Syndrome

- Highest incidence between 6 months & 2 years
- Painful symmetric swelling of the dorsum of hands & feet due to ischemia
- May be unilateral
- Occurs of 50-75% of Hb S patients
- Can be confused with osteomyelitis
- Treatment
  - Hydration
  - Pain control
  - Reassurance

Priapism

- Painful persistent erection of the penis due to sickling in the corpora cavernosa
- Usually occurs between 5-35 years of age
- Usually preceded by multiple short episodes
- Precipitated by:
  - Infection
  - Intercourse \ masterbation
  - Nocturnal erection with early morning awakening
- Treatment
  - Usual sickle cell crisis measures
  - Exchange transfusion
  - Surgical shunting
  - Impotence occurs in 1\3 to 1\2

Retinopathy

- Chronic ischemia leads to neovascularization & increased risk of:
  - Vitreous hemorrhage
  - Retinal detachment
  - Blindness
- Hb SC and Hb S thal are at increased risk

Nephropathy

- Hyposthenuria leads to:
  - Inability to concentrate urine
  - Nocturia & enuresis
  - Tendency toward dehydration
  - Reduced resistance to urosepsis
- Common Renal Manifestations:
  - Hematuria
  - Papillary necrosis
  - Proteinuria
  - Renal tubular acidosis
  - Nephrotic syndrome
  - Glomerulosclerosis
  - Renal insufficiency

Pulmonary

- Common Symptoms of:
  - Pneumonia
  - Acute chest syndrome
  - Fat embolization
  - Rib Infarctions
- Older Patients:
  - Chronic restrictive lung disease
  - Pulmonary hypertension
  - Cor pulmonale (pulmonary hypertension)

Acute Chest Syndrome

- Etiology
  - Sickling in the pulmonary arteries leading to pulmonary infarction.
  - May also be caused by pneumonia especially in children
- Symptoms
  - Acute pleuritic chest pain, hemoptysis, fever, leukocytosis, + / - pulmonary infiltrates
- Diagnostic Test
  - V / Q Scan
- Treatment
  - Admission
  - Treat hypoxia and respiratory distress
  - Empiric antibiotics
  - Exchange transfusion for severe episodes

Leg Ulcers

- Occur in 25-50% during lifetime
- Related to vascular stasis
- Treatment is common to other types vascular stasis
- Avoid starting IV in lower extremities

Abdominal Pain

- Increased Incidence
  - Cholecystitis
  - Peptic ulcer
  - Pyelonephritis
  - Bowel infarction
- Almost Exclusive
  - Splenic \ hepatic sequestration
- Typical Pain Episode

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Clinical Case

A 28 year old black male with known Hb SS disease presents to the ED with "sickle cell crisis". This is his 52 visit to the ED in the last 6 months. An astute intern (with nothing better to do) reviews the old chart and adds up the amount of Demerol this patient has received in the last 6 months.

Demerol: 16,450mg
Hours in 6 months: 4380
Essential Drip Rate: 4mg\hour

What would you do now?

Sickle Cell Pain Episode "Crisis"

- Most common acute problem
- "A self-limited episode of diffuse, reversible pain often occurring in the extremities, back, chest, & abdomen."
- Severity ranges from mild transient 5 minute attacks to excruciating pain lasting days to weeks requiring hospitalization.
- Caused by the inflammatory response of bone or marrow necrosis, muscle or bowel ischemia secondary to sludging of sickled RBC's
- Patient may have a fear of death but never a cause of mortality unless caused by another underlying problem.

Sickle Cell Pain Episode "Crisis"
Diagnosis

The is no characteristic clinical finding which can make the diagnosis or define the severity of a pain episode, therefore the patient's assessment must usually be accepted.

Sickle Cell Pain Episode "Crisis"

- Variable Frequency
  - Hemoglobin phenotype
  - Physical condition
  - Smoking
- Precipitating Factors
  - Increased physical or psychological stress
  - Fever
  - Dehydration
  - Overexertion
  - Rapid temperature change
  - Anger
  - Often occur without antecedent cause

Sickle Cell Pain Control
Serotonin Enhancement

- Purpose
  - Adjuvant to analgesics
  - Decreases the perception of pain
  - May be beneficial in treatment of chronic pain
- Types
  - Tricyclic antidepressants
  - L-tryptophan

Sickle Cell Pain Control
Peripheral Pain Reduction

- Types
  - Anti-inflammatory: Aspirin & NSAIDs
  - Local heat
  - Immobilization

Sickle Cell Pain Control

Enkephalins & Endorphins Endogenous Opioids

- Direct Stimulation
  - Neuronal activation from higher brain centers
- Indirect Stimulation
  - Biofeedback relaxation techniques
  - Transcutaneous electrical nerve stimulation (TENS)
  - Acupuncture

Sickle Cell Pain Control

Exogenous Opioids

- Mechanism
  - Binding of opiate receptors in the brain
  - Similar to endogenous opioids
- Side Effects
  - Somnolence
  - Respiratory depression
  - Nausea \ vomiting
  - Hypotension
  - Constipation
  - Increased bladder tone \ urinary retention
  - Local irritation at injection site
  - Decreased seizure threshold
  - Histamine release (morphine)
- Agonist-Antagonist
  - May precipitate withdrawal syndrome in narcotic conditioned patients
  - Buprenex (buprenorphine)
  - Nubain (nalbuphine)

Management of Pain Episode

Clinical Evaluation to Exclude

- Correctable Precipitants
  - Infection
  - Dehydration
  - Acidosis from any cause
  - Emotional stress
  - Extreme temperature exposure
  - Alcohol or drug abuse
- Life-threatening complications
- Causes of pain unrelated to sickle cell

Management of Pain Episode
Precipitating Complication

- Uncharacteristic pain for the individual
- New physical finding
- Significant changes in laboratory tests
  - Significant decrease in hemoglobin
  - Markedly decrease reticulocyte count
  - Markedly increase WBC

Treatment of Pain Episode

- Oral or IV hydration
  - ½ NS at 250cc\hr (in absence of CHF, renal failure, hyponatremia)
  - Change to D_5 ½ NS or ½ NS at slower rate after 1st liter.
- Bed rest
- Treatment of underlying illness

Treatment of Pain Episode
Analgesics

- Mild Episode
  - Aspirin, Acetaminophen +\- Codeine, hydrocodone, NSAID
  - Usually these have already been tried

Treatment of Pain Episode
Analgesics

- Moderate to Severe Episode
  - Oral or Parenteral NSAID (Toradol 30mg IV or 60mg IM)
  - Narcotics (sustained release)
    Initial doses depend on patient's prior narcotic use & subsequent doses should be titrated according to response to initial doses.
    - Morphine 0.1-.02 mg/kg (max 15mg) IM\IV
    - Levo-Dromoran 2mg IM\Sq\PO (4-6mg if heavy narcotic user)
    - Methadone 10mg IM
    - *Nubain 0.3 mg/kg (max 20mg) IM\IV
    - *Buprenex 0.3-0.6 mg initially IM\IV

    **Agonist-Antagonist: Do not use if physically dependent on narcotics.*
  - Adjuvants
    - Vistaril 25mg IM
    - Phenergan 25mg IV\IM
    - Inapsine (droperidol) 2.5mg IV\IM

Treatment of Pain Episode
Outpatient

- If treatment can be given for up to 8 hours in an outpatient setting > 80% will respond sufficiently to allow outpatient management.
- Practical consideration may require a less lengthy ED treatment course.

Treatment of Pain Episode
Outpatient

- Continued Hydration
- Rest
- Medication (48 Hour Supply)
  - NSAID
  - Codeine\APAP (Tylenol #3 or #4)
  - Hydrocodone
  - Oxycodone (OxyContin)
  - MS Contin or MS Liquid
  - Levo-Dromoran
  - Methadone
  - *Stadol NS

Treatment of Pain Episode

Admission Criteria

- Pain persists more than 8 hours in an outpatient setting.
- Failure of outpatient therapy with return within 48 hrs.
- Complications Present
  - Infection
    - WBC > 20k &\ or significant left shift
    - Fever > 101° F
    - Documented infection (i.e. pneumonia)
  - Significant hypoxia or acidosis
  - Pregnancy
  - Cardiac decompensation
  - Priapism
  - Thrombo-embolic event to lungs or CNS
  - Aplastic or hyperhemolytic crisis (acute fall of Hgb > 1gm\dl)
  - Hepatic syndrome or cholecystitis

Treatment of Pain Episode

Inpatient

- IV hydration
  - D_5 ½ NS or ½ NS at 5cc\kg\hr (200-300 cc\hr in adults)
  - Oral hydration is acceptable if no IV site and not nauseated
- Bed rest
- Treatment of underlying illness

Treatment of Pain Episode

Inpatient Analgesics

- Sustained Release:
  - Morphine, Levo-Dromoran, Methadone
- FIXED Schedule of Administration - NOT PRN
  - All that hurts is not sickle cell pain!
  - Based on duration of action
  - PCA pump or fixed nursing schedule
  - Periodic adjustment of dosage (not frequency of administration) based on level of pain control & sedation.
- PRN IV Administration NOT Recommended
  - High acute blood levels
  - Excessive euphoria
  - Respiratory depression
- Discharge
  - Provide less than 48 hour supply of narcotic medication
  - Sustained release

Chronic Sickle Cell Pain

Prolonged & Constant Pain

- Causes
  - Bone infarction
  - Sickle Arthritis
  - Aseptic necrosis of femur or humerus
- Treatment
  - Hydroxyurea (decreased pain crisis & admits by 50%
  - NSAIDs with renal sparing properties (Relafen)
  - Tricyclic antidepressants
  - TENS unit
  - Biofeedback, relaxation techniques, self-hypnosis
  - Occupational & physical therapy
  - Education & psychological support

Chronic Sickle Cell Pain

Prolonged & Constant Pain

- Opiate analgesics
  - Prevent inappropriate continuous use of opiates
  - In general narcotics should be reserved for acute pain crisis and not used for chronic pain
- Severe & Moderate Pain
  - Sustained release preparations
  - MS Contin, Levo-Dromoran, Methadone, OxyContin
  - Avoid prescribing Codeine \ APAP (Tylenol #3 or #4) hydrocodone (Vicodin) or oxycodone (Percocet)

Clinical Case

A 24 year old black female with known Hb SS disease is found unconscious with depressed respirations.

VS: HR-135 RR-4 BP-100 \ 50

What do you do next?

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Clinical Case

A 24 year old black female (same patient as before) with known Hb SS disease was admitted to the hospital for "acute pain crisis". 24 hours after admission she was found confused & hallucinating. She subsequently had a grand mal seizure.

VS: HR-145 RR-32 BP-130\95

What happened?

Demerol (Meperidine)

- A drug designed for failure in treatment of chronic recurring pain syndromes.
- Short acting
- Marked euphoria
- Highly psychologically addicting
- Often the "drug of choice"
- Significant toxicity in large & chronic doses
  - Nor-meperidine Toxicity
  - Serotonin Syndrome

Serotonin Syndrome

- Similar to neuroleptic malignant syndrome
  - Fever, muscle rigidity, mental status changes, seizures
- Associated with:
  - SSRIs: Prozac, Paxil, Zoloft, Serzone, etc.
  - Pondimin, Redux
  - Meperidine (esp. with MAO Inhibitors, eg. Nardil)
  - Antihistamines
  - Sinemet

Sickle Cell & Drug Abuse

- Does not predispose or provide protection from abuse.
- The combination however poses significant challenges
  - Compromises ability to diagnose & treat complications
  - Pain crisis less responsive to treatment
  - Addiction treatment complicated by true underlying painful disease
- Pattern of Addiction
  - Frequency & duration of narcotic use
  - Drug seeking behavior \ Anxiety regarding being without
  - Evidence of psychological & physical dependence
  - Disruption of normal functioning
  - Fixation on narcotic of choice
  - Continued use despite adverse reactions (meperidine toxicity)
  - Multiple allergies to alternative narcotics

Sickle Cell & Drug Abuse

Treatment

- Detoxification over 6 weeks or less with gradual reduction in narcotic use
- Maintain narcotic free status as outpatient
- If outpatient narcotic is necessary, it is delivered through an established drug dependency unit
- Pain Crisis:
  - Evaluated & treated in a hospital setting only
  - No outpatient narcotics are ever provided
  - Agonist-Antagonist narcotics for acute pain episodes for short durations only.
  - Patient's drug of choice should NEVER be used again for pain crisis

Sickle Cell & Drug Abuse

Treatment

- Compliance to with treatment after detoxification monitored by random drug screens
- Supportive Services:
  - Counseling
  - Vocational rehabilitation
  - Narcotics anonymous
- Failure to maintain drug free status carries significant reprisals

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Clinical Case

A 28 year old black male with known Hb SS disease is admitted to the hospital for "acute pain crisis". Having just attended an excellent lecture on SS pain crisis management the doctor orders a regimen of Levo-Dromoran.

During the weekend the nurse (who has not attended the above mentioned lecture) calls the covering doctor multiple times relaying the patient's demands for Demerol.

This doctor changes the order to Demerol 50mg IV q2h prn.

What do you think happens?

Sickle Cell Management Protocol for the "Difficult" Patient

Page 1 of 4

Based on: Problem Oriented Management of Sickle Syndromes

143 Page Handbook By:

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Sickle Cell Management Protocol for the "Difficult" Patient

Page 2 of 4

- Chronic transfusion program until:
  - Drug treatment completed
  - Narcotic free for more than 3-6 months
  - Re-initiation any time > one "pain crisis" in a 30 day period
- Narcotic detoxification
  - Initial in-patient followed by out-patient drug treatment
  - No out-patient narcotics
  - Routine drug testing to assure compliance
- "Pain crisis" treatment is limited to:
  - One ED & PCP
  - Responsible for tracking each crisis

Sickle Cell Management Protocol for the "Difficult" Patient

Page 3 of 4

- Patient cooperation
  - Fully advised of the treatment program
  - Give permission for notification to all local ED's
  - Sign a contract of intended compliance
  - Must agree to the proscribed treatment
  - Many sickle cell patient who arrive at this stage will not likely agree initially, if at all

Sickle Cell Management Protocol for the "Difficult" Patient

Page 4 of 4

- The uncooperative patient: *Treatment by committee*
  - Similar to an institutional review board
  - Review & address the medical history
  - Decide if the patient should ever receive narcotics even in an in-patient setting unless compliant with a treatment program
- The pattern is usually clear:
  - Over utilization of resources
  - History of illegal forgery of prescriptions
  - Obvious narcotic dependence
- Continued narcotic administration without compliance with a rational proscribed treatment program is not in the patient's best interest

Multi-Disciplinary Approach to Sickle Cell Patient Management

- Sickle Cell Patient Management Team
- Standardized approach to treatment
  - Inpatient and outpatient
- Education
  - house staff, attending staff, nursing, patients
- Evaluate
  - Satisfaction (patient & provider)
  - ED visits
  - Inpatient admissions
  - Length of stay

Integrated Approach to the Emergency Management of Sickle Cell Anemia Pain Crisis

ACEP Scientific Assembly - Wednesday, October 13, 1999 - (WE172)

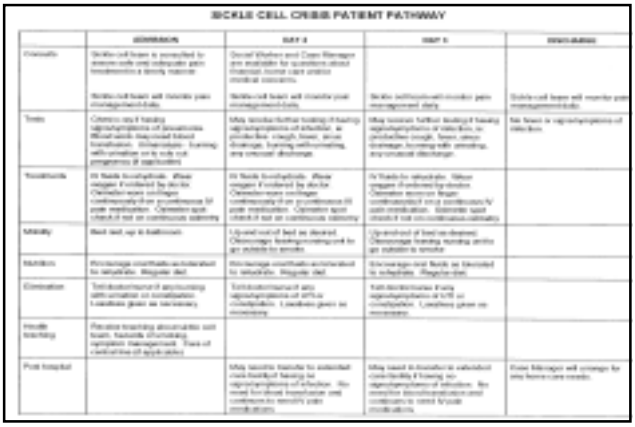
Todd B. Taylor, MD, FACEP

Management Team

- Monthly Meetings
  - ED: physician & nursing
  - Social work
  - Pharmacy
  - "Pain management" nurses (oncology)
  - Case management
  - Quality improvement director
  - Hematologist
- Inpatient consults available 24/7
- Assess every sickle cell admission

Standardized Treatment Approach

- Inpatient\ED
  - Individualized patient profiles
  - Standardized admission orders
  - Treatment pathway & goals (discharge <4 days)
- Every admission reviewed by a team member
- Problems & future plans reviewed monthly



SICKLE CELL CRISIS PATIENT PATHWAY

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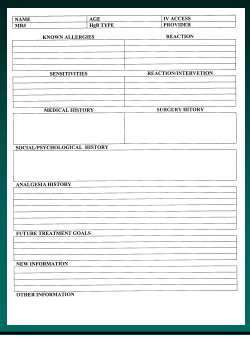
Sickle Cell Patient Profile



NAME | AGE | IV ACCESS
MR# | Hgb TYPE | PROVIDER

KNOWN ALLERGIES | REACTION

SENSITIVITIES | REACTION/INTERVETION

MEDICAL HISTORY | SURGERY HISTORY

SOCIAL/PSYCHOLOGICAL HISTORY

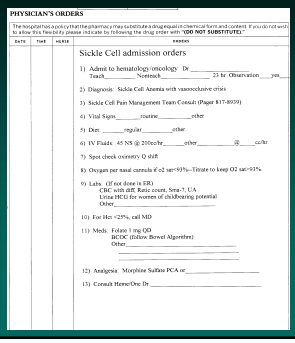
ANALGESIA HISTORY

FUTURE TREATMENT GOALS

NEW INFORMATION

OTHER INFORMATION

Standardized Order Sheet



PHYSICIAN'S ORDERS

The hospital has a policy that the pharmacy may substitute a drug equal in chemical form and content. If you do not want it to allow this flexibility please indicate by following the drug order with "**DO NOT SUBSTITUTE**."

DATE | TIME | NURSE | ORDERS

Sickle Cell admission orders

1) Admit to hematology/oncology Dr._____
Teach_____Nonteach_____23 hr Observation____yes____

2) Diagnosis: Sickle Cell Anemia with vasoocclusive crisis

3) Sickle Cell Pain Management Team Consult (Pager 817-8939)

4) Vital Signs_____routine_____other

5) Diet: _____regular/_____other

6) IV Fluids: 45 NS @ 200cc/hr_____other_____@_____cc/hr

7) Spot check oximetry Q shift

8) Oxygen per nasal cannula if o2 sat<93%--Titrate to keep O2 sat>93%

9) Labs: (If not done in ER)
CBC with diff, Retic count, Sma-7, UA
Urine HCG for women of childbearing potential
Other_____

10) For Hct <25%, call MD

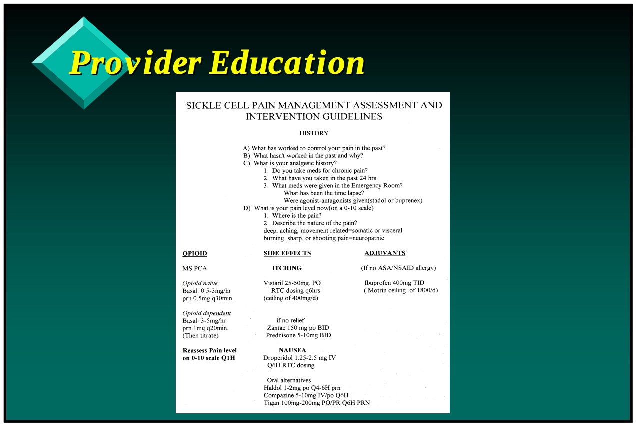
11) Meds: Folate 1 mg QD
BCOC (follow Bowel Algorithm)
Other _____
_____
_____

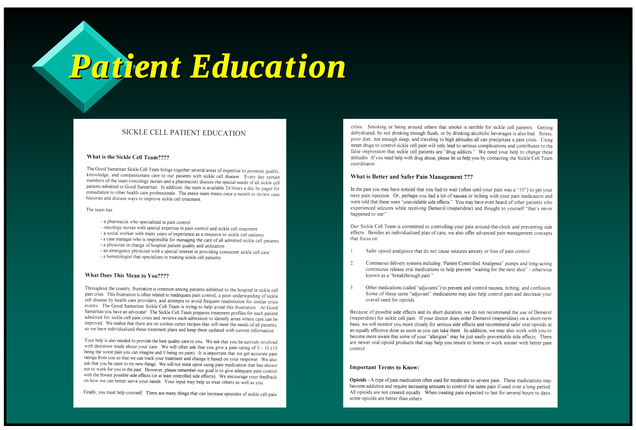
12) Analgesia: Morphine Sulfate PCA or_____

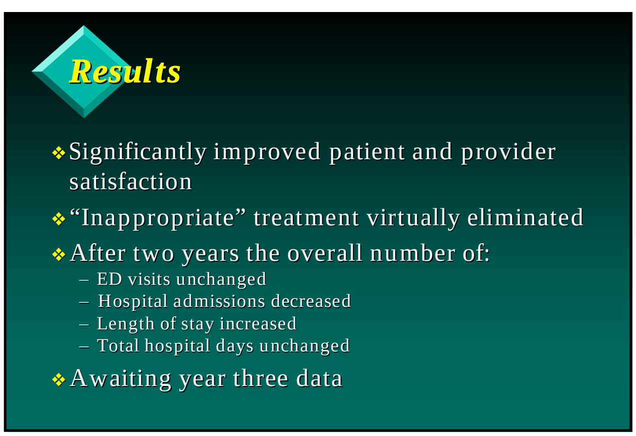
13) Consult Heme/Onc Dr._____

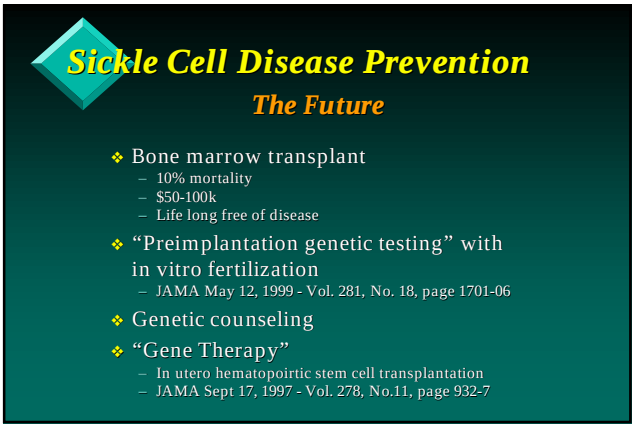
Education

- Provider
  - House staff
  - Attending staff
  - Nursing
  - Health plan
- Patient & Family


Provider Education
SICKLE CELL PAIN MANAGEMENT ASSESSMENT AND INTERVENTION GUIDELINES
HISTORY
A) What has worked to control your pain in the past?
B) What hasn't worked in the past and why?
C) What is your analgesic history?
1. Do you take meds for chronic pain?
2. What have you taken in the past 24 hrs.
3. What meds were given in the Emergency Room?
What has been the time lapse?
Were agonist-antagonists given(stadol or buprenex)
D) What is your pain level now(on a 0-10 scale)
1. Where is the pain?
2. Describe the nature of the pain?
deep, aching, movement related=somatic or visceral
burning, sharp, or shooting pain=neuropathic
OPIOID
MS PCA
Opioid naive
Basal: 0.5-3mg/hr
prn 0.5mg q30min.
Opioid dependent
Basal: 3-5mg/hr
prn 1mg q20min.
(Then titrate)
Reassess Pain level on 0-10 scale Q1H
SIDE EFFECTS
ITCHING
Vistaril 25-50mg. PO
RTC dosing q6hrs
(ceiling of 400mg/d)
if no relief
Zantac 150 mg po BID
Prednisone 5-10mg BID
NAUSEA
Droperidol 1.25-2.5 mg IV
Q6H RTC dosing
Oral alternatives
Haldol 1-2mg po Q4-6H prn
Compazine 5-10mg IV/po Q6H
Tigan 100mg-200mg PO/PR Q6H PRN
ADJUVANTS
(If no ASA/NSAID allergy)
Ibuprofen 400mg TID
(Motrin ceiling of 1800/d)


Patient Education
SICKLE CELL PATIENT EDUCATION
What is the Sickle Cell Team????
What Does This Mean to You????
What is Better and Safer Pain Management ???
Important Terms to Know:


Results
Significantly improved patient and provider satisfaction
"Inappropriate" treatment virtually eliminated
After two years the overall number of:
– ED visits unchanged
– Hospital admissions decreased
– Length of stay increased
– Total hospital days unchanged
Awaiting year three data


Sickle Cell Disease Prevention
The Future
Bone marrow transplant
10% mortality
$50-100k
Life long free of disease
"Preimplantation genetic testing" with in vitro fertilization
JAMA May 12, 1999 - Vol. 281, No. 18, page 1701-06
Genetic counseling
"Gene Therapy"
In utero hematopoirtic stem cell transplantation
JAMA Sept 17, 1997 - Vol. 278, No.11, page 932-7

Sickle Cell Patient Profile

NAME	AGE	IV ACCESS
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