Objectives

1. List two SCD Clinical Syndromes.
2. Differentiate characteristics of SCD pain.
3. Describe the phases vaso-occlusive pain episodes.
4. Describe factors affecting SCD HRQOL.
5. Describe 2 goals of SCD Pain Assessment.
6. Describe evidenced based approach to SCD Pain Management

Sickle Cell Disease

- Genetic disorder
  - Autosomal dominant inheritance
  - Persons of African, Caribbean, Mediterranean descent
  - Complex phenotypes and haplotypes

- Genotypes
  - HB AS
  - HB SS
  - HB SC
  - Beta Thalassemia’s
  - Beta +
  - Beta –
  - Combinations possible
Sickle Cell Pain Characteristics

- Acute (vaso-occlusive pain episodes – ‘crises’)
- Chronic (Persistent)
- Nociceptive
- Neuropathic
- Mixed
- (nociceptive-neuropathic-chronic-acute)

Sickle Cell Pain

- Unpredictable
- Uncontrollable
  - Onset
  - Frequency
  - Duration
  - Cause
- Severe pain
- Intra and inter individual variability

- Avascular Necrosis
- Bony infarctions
- Osteoarthritis (Septic Arthritis/Osteomyelitis)
- Unilateral or Bilateral
- Chronic Pain Syndrome
- Neuropathic Pain Syndromes
  - Femoral head
  - Humeral head
  - Patella
  - Hips
  - Vertebrae
Laboratory Correlates of SCD Clinical Severity

- **Decreased Hemoglobin**
  - cardiomyopathies
  - strokes
    - Silent
    - Infarctive
    - hemorrhagic
  - spleen
    - sequestration crises
    - Autoinfects

- **Decreased Hgb F**
  - vaso-occlusive pain frequency
  - spleen
    - sequestration crises
    - Autoinfect
  - leg ulcers
  - acute chest syndrome

- **Increased Hematocrit**
  - vaso-occlusive pain frequency
  - osteonecrosis femoral head

- **Increased WBC**
  - silent infacts
  - hemorrhagic stroke

- **Increased vaso-occlusive pain frequency**
  - premature death
Healthcare Professional Barriers in Sickle Cell Disease Pain Management

Lack of pain assessment skills  
Failure to accept patients report of pain  
Personal opinions and beliefs  
Fear of overdosing/causing addiction  
Lack of understanding/communicating about pain  
Lack of SCD Knowledge

Pseudoaddiction

Direct consequence of inadequate pain management.

- Clinical interaction-describes patient behavior that may occur when pain is under treated.
- Leads to behaviors such as: ‘drug seeking’, clock-watching, and deception in an effort to obtain relief of pain.
- Behavior ceases when adequate pain relief provided.

American Medical Association - 2003, Weissman, Haddock, Pain

www.ampainsoc.org/pub/sc.htm

American Pain Society August, 1999
APS (1999) SCD Pain Management Guidelines

**VOE Acute Pain Episode Assessment**

- Typical Sickle Cell Pain?
  - NO: Determine Cause
  - YES: Continue Sickle Cell Pain Management

- Nociceptive Pain
  - Sickle Cell Pain
  - Vasculitis
  - Infections
  - Inflammation
  - Migraine
  - Malignancy
  - Post-surgery

- Neuropathic Pain
  - Sickle Cell Pain
  - Demyelination
  - Nerve Injury

- Other Causes
  - Acute Gout
  - Acute Rheumatic Arthritis
  - Arthritis
  - Cholecystitis
  - Cholangitis
  - Dehydration
  - Kidney Stones
  - Migraine
  - Muscle spasticity
  - Myocardial Infarction
  - Nephrolithiasis
  - Pancreatitis
  - Peritonitis
  - Priapism
  - Aplastic Anemia
  - Necrotizing Fasciitis
  - Sequestration

**Opioids—Morphine is Gold Standard**

- Loading Dose
  - Morphine: <50 kg
    - 0.1 - 0.15 mg/kg
  - Morphine: >50 kg
    - 5 - 10 mg dose
  - Dilaudid: <50 kg
    - 0.015 - 0.020 mg/kg
  - Dilaudid: >50 kg
    - 1.5 mg dose

**Side Effects**

- Pruritus
- Nausea/Vomiting
- Sedation/Constipation

**Chronic Opioid Therapy?**

- Continue during inpatient admission
- Tolerance/Dependence
- Pseudoaddiction

**Obtain Tx History**

- What works BEST for THIS patient?
  - PCA
  - Epidural

**REFERENCES UPON REQUEST**

- Intrahepatic/Hepatic Sickling
- Osteoarthritis
- Joint Effusions
- Avascular Necrosis
- Sickle Arthritis
- Other
- Pancreatitis
- Cholelithiasis
- Constipation
- Bowel Obstruction
- Priapism
- Bronchitis/Pneumonia
- Acute Chest Syndrome
- Splenic Sequestration
- Liver Sequestration
Questions and Reference List

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