Opioid Prescribing for Adults with Sickle Cell Disease

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Dr. Hagar, Disclosures

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Learning Objectives

- Overview of Uniqueness of Sickle Cell Pains
- Know the critical role of the doctor-patient relationship in pain management
- Know how to approach new sickle cell patients about their pain medication requirements
- Know how to evaluate efficacy of treatment
- Know when to decrease or stop opioids

Overview of Uniqueness of Sickle Cell Pain

Provider Concerns

- Worry about whether chronic pain management is helping with analgesia or facilitating drug addiction
- Concern about with patients with co-occurring disorders
- Worry about assessing the effects of their treatments
- Concern about a lack of a standard approach

Provider Concerns

- These concerns are typical for treating any chronic pain
- Sickle cell patients likely have less addiction than the background population
- Basic knowledge and some experience with treating sickle cell patients will give the provider the skills and knowledge to address these concerns.
- Provider attitudes improve when they learn that they have the ability to help

Sickle Cell Pain

- Recognized as a unique pain syndrome
  - Pathophysiology involves neuroinflammatory components, central pain pathways, and psychosocial modifiers
- Understudied
  - Only about 100,000 patients in US
  - Sickle Cell Disease is officially classified as a rare disease

Sickle Cell Pain

- Multiple Components
  - Neuroinflammatory and cytokine driven pain
  - Nociceptive and neuropathic components
  - Anticipatory pain
  - Opioid responsive components
  - Peripheral and Central pain components
  - Psychosocial pain modifiers

Pain Often is the Earliest Symptom of a Life Threatening Event

Patients often have repeated episodes of typical pain, pain, pain, then suddenly have a life threatening event such as acute chest syndrome, mulitorgan failure, splenic or hepatic sequestration, and infections

Know the Critical Role of the Doctor-patient Relationship in Pain Management

Approach to an Adult Office Patient with Sickle Cell Pain

- Key Points to keep in Mind
  - Patients perception of providers concern is most important “adjuvant” for treatment
    - You don’t have to agree with the patient, but the patient needs to feel that you have listened, understood, and considered their concerns
  - Your goal is not “perfect” pain control, but a level of functional pain that can be evaluated and modified

Patient Approach Framework

- Patients will have different patterns of pain and responses
  - These are often stable for the individual
  - Ask the patient what their pain is usually like
  - For acute pain, ask if this is similar to past episodes
- *There is no one way to predict who is in pain*

Patient Approach Framework

- Screen for the often co-existing mental illness issues
- Sign up for access to your state Prescription Drug Monitoring Program (48 states have such programs, 2 are pending) www.pmpalliance.org
- Check PDMP prior to starting to prescribe scheduled drugs and at regular intervals (e.g.: q 3-4 months) thereafter
- Inform the patient of regular, but random urine and blood testing for licit and illicit drugs.

Know How to Evaluate Efficacy of Treatment

Safe Practice

- In 2010, there were 107,000 visits for opioid exposure in patients with Sickle Cell Disease, with 27,500 requiring hospital admission
- Concurrent use of benzodiazepines are biggest risk facture
- Children often exposed through unsecured home opioid supplies
- Paradoxically, patients with long history of exposure to opioids remain at risk of respiratory depression

Patient Approach Framework

- Sickle Cell has become a Chronic Disease
  - We now have the highest number of adults with sickle cell disease in human history , so little cumulative experience with much of what patients are experiencing
- Most adults will have some component of centralized pain syndrome
  - Pain will be out of proportion to signs
  - Patients will have allodynia and hyperesthesia

Note That:

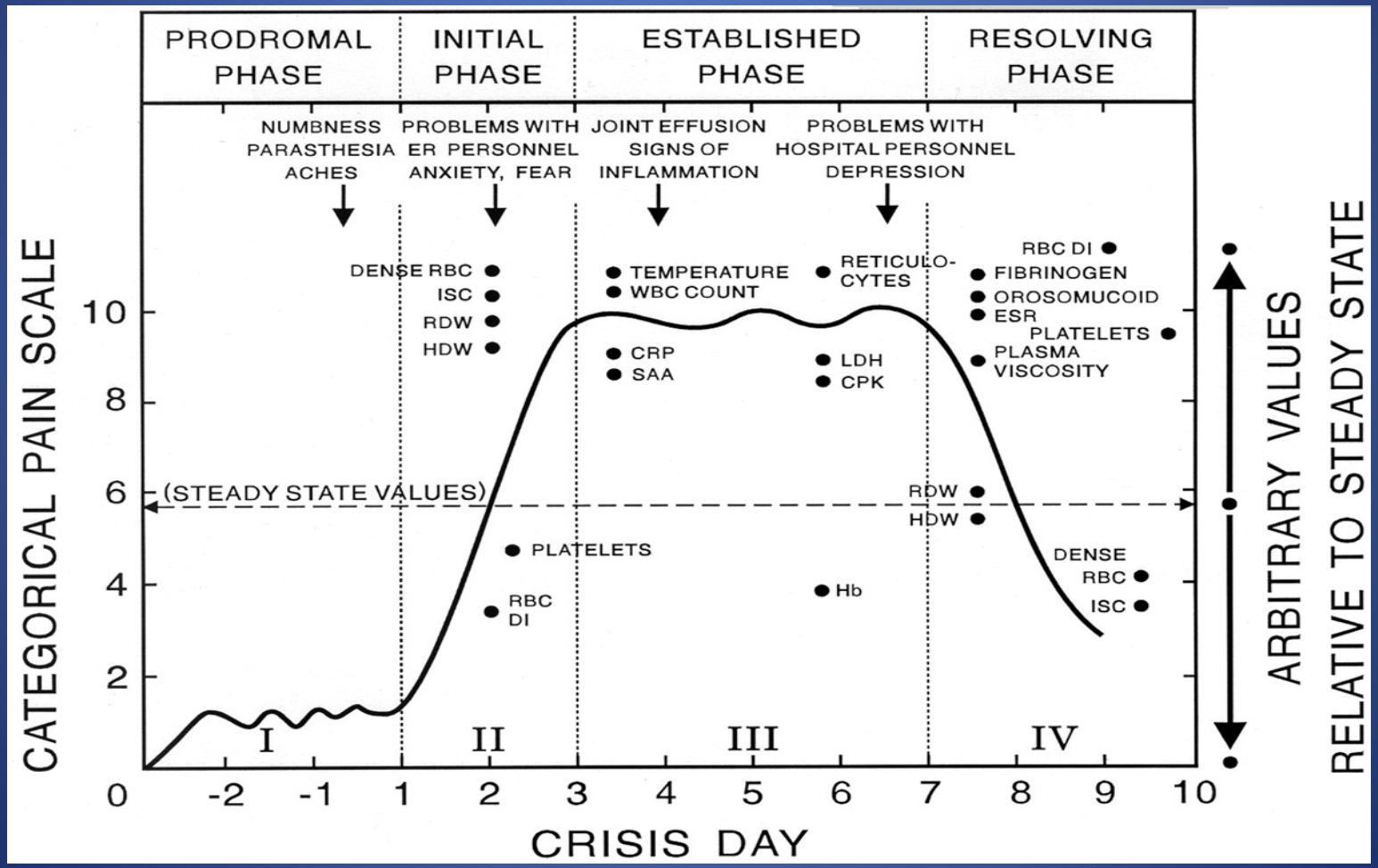
- Most patients deal with pain of similar severity and duration at home
- Most patients have had pain for days before arriving at office or ED for treatment
- Patients are usually sleep deprived and irritable from unrelenting pain when they present with acutely
- Often the only relief for pain not responsive to opioids is drowsiness

Note That Also:

- The best time to treat pain is the prodromal phase
- Most adults know what works for them
- Until there is a reason not to, believe the patient about what they need. They have lived with their pain for a lifetime.
- Anticipatory pain is a driver of prolonged pain (see next slides)

Phases of Pain Crisis

If you can intervene in the prodromal phase, you can stop or decrease subsequent phases. Learning an individual's symptoms and lab responses to pain can predict, for that patient, whether they are in crisis.



Ballas, Hematology, 2007

Anticipatory Pains

- Fear of severe pain returning causes viscious cycle of stress-induced pain
- Increase in stress markers show physiologic neuroactivation
- Knowing that someone is going to listen to them to reassess pain and to adjust their treatment decreases total opioid use and time in pain

Know How to Approach New Sickle Cell Patients About Their Pain Medication Requirements

Approach to New Patient with Sickle Cell Disease

- Some organized time at initial interview will dramatically improve long term care
- Listen to patient's story about their pain
- Most patients have horror stories about how physicians didn't listen to them
- Review usual pain course, what worked, and if some treatment *didn't* work, why not. (see next slide)

Sample Pain History

- Mr. A's sickle pain is different from his chronic low back pains. The sickle pains are throbbing pains that begin in his mid-back and radiate to arms. He knows his crisis is becoming severe when the pains radiate to his legs. He tries warm baths, fluids, and hydromorphone. When his hydromorphone doesn't "take the edge" off his pains, he comes to the ED.

Approach to New Adult Sickle Cell Patient with Pain

- Is this typical pain?
  - Patients differ in descriptions of usual pain, but are remarkably consistent by individual
  - If not, then look for other causes
- Ask the patient what has worked in the past
  - Document patient response
- Be aggressive first 48 hours
  - Decreases central pain syndromes
  - Breaks pain cycle sooner “Gets ahead of the pain”

Know When to Decrease or Stop Opioids

Goals for Pain Control in Clinic

- Look at number of ED visits and hospitalizations
- Use level of function as best indicator of success
- Goal is NOT total pain relief (we wish this were possible) but the least amount of effective medications to allow functioning
- If patient is staying out of the ED and hospital and has a good level of function, don't worry about total dose

Doses

- Be aware of prior opioid doses
  - Some patients so seldom have pain that even low dose opioids may depress respiration
  - 25% of adults with sickle cell have daily pain
  - Current approach is to be aggressive over the first 48 hours, both to get ahead of the pain and to decrease anxiety, fear, and anticipatory pain

Doses of Opioids

- Doses vary widely between patients
- Goal is function
  - Most patients require doses of 30 mg of morphine equivalents or less
  - A few require higher doses, some much higher doses (e.g., one patient placed on 600 mg morphine thrice daily plus breakthrough medications went from weekly admissions to no admissions for five years)
  - Regular assessments needed for adjustments

Typical IV Doses For ED and Hospital Use

- Hydromorphone – 2 to 4 mg IV q2 -4 hours, hold for sedation
- Morphine sulfate – 4 to 8 mg IV q1 – 3 hours, hold for sedation
- Often lorazepam 1 mg IV is added to decrease anxiety
- Often ketorolac 30 mg IV q6h is added to help with inflammatory component

Adjuvants Are Important

- Fluids (0.5 M normal saline), no potassium (damages small veins) 120 ml/hr; higher fluid rates lead to pulmonary edema
- Ketorolac or ibuprofen helps any inflammatory component
- Lorazepam helps anxiety, spasm, and anticipatory pain

IMPORTANT

- Most important principle is regular, rapid assessment and treatment adjustment
- Opioids can be safely given intravenously every 10 to 20 minutes if reassessed
- Patients often sleep when pain breaks
- Just having reassessments and medication adjustments reduces pain

Patient Relations

- Let the patient know that you believe they are in pain
- Let the patient know you will re-evaluate the effectiveness of medications administered
- Give the patient as much choice as medically reasonable
- Patients are not at their best when in severe pain

When to Decrease Medication

- No improvement in functioning has been achieved
- No decrease in ED visits or hospitalizations
- Reports little improvement when taking medication or medication works only for a short time

In Clinic

- Need to find stable dosing that allows functioning with fewest side effects
- Need frequent clinic appointments to assess whether or not the pain medication is adequate or needs adjustment
- Once pain pattern and response to treatment is explored, visits can be spaced further out

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