

Emergency Medicine Vaso-occlusive Crises



Board Questions

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Question 1



- ☞ A 16yo HbSS male, with a history of two prior episodes of priapism presents to the Emergency Department with a painful erection which started less than 2hours ago. What is the next best step in his management?
- a) Aspiration and irrigation of the corpus cavernosum
 - b) Instillation of phenylephrine into the corpus cavernosum
 - c) IVF and morphine
 - d) Exchange Transfusion
 - e) Winter Procedure

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Priapism



- ❧ Priapism is an abnormal and painful erection of the penis that is involuntary and not relieved by ejaculation.
- ❧ Due to stasis and low outflow secondary to vaso-occlusion within the venules and sinuses of the corpus cavernosum
- ❧ True urologic emergency.
- ❧ Stasis of deoxygenated blood can cause fibrotic changes within the cavernosum within 12-24 hours and can lead to permanent impotence.

Management of Priapism



- ❧ Less than 2hours
 - ❧ Drink extra fluids, use oral analgesics, exercise, and attempt to urinate.
 - ❧ Report to the ED for i.v. hydration and morphine to treat sickling crisis
 - ❧ Oral terbutaline or pseudoephedrine

Management of Priapism



- ❧ Over 2hours
 - ❧ Aspiration of the corpus cavernosum
 - ❧ Often aspiration of small amounts of blood (20-40 cc) from only one side will relieve symptoms.
 - ❧ If unsuccessful, direct injection of phenylephrine or epinephrine is performed.



Management of Priapism



- ❧ Surgical management
 - ❧ Considered when Priapism lasts beyond 12hours and after aspiration and instillation have failed
 - ❧ Winter Procedure involves shunting blood into corpus spongiosum to relieve vascular occlusion

Role of Blood transfusion



- ❧ Controversial
- ❧ Associated with acute neurological events due to rapid elevation of Hb and release of procoagulant factors
- ❧ Exchange Transfusion to Hb <10g/dl preferred
- ❧ Exchange Transfusion as sole tx delays detumescence to 24 -48hrs

References



- ❧ www.uptodate.com - Diagnosis and management of priapism in sickle cell disease
- ❧ Medscape Pediatrics - Treatment of Priapism in Pediatric Patients With Sickle Cell: Adrenergic Agents
- ❧ Bruno D, Wigfall DR, Zimmerman SA et al. Genitourinary complications of sickle cell disease. *J Urol.* 2001; 166:803-11.

Question 2



- ☞ An 13yo HbSS female presents to the ED complaining of bilateral knee pain rated 6/10. Her last painful crisis was over one year ago. Which of the following statements regarding the appropriate management of her pain is correct?
- a) NSAIDS are ineffective in the management of moderate pain
 - b) Opioids are only administered for patients with severe intractable pain
 - c) Assessment of iv drug effect should be made every 15-30minutes
 - d) Meperidine is safe to use in patients with impaired renal function

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Explanation – Re-evaluation



- ☞ Evaluate the response to therapy 15 to 30 minutes after each dose by assessing pain intensity, relief, mood, and sedation level.
- ☞ Record the pain assessment and reassessments, along with the patient's other vital signs, in the patient's chart and/or on a bedside flowsheet

Explanation - NSAIDs



- ❧ Mild-to-moderate pain in children generally is managed with NSAIDs or acetaminophen
- ❧ Most NSAIDs are given only orally, except for ketorolac, which can be used orally or parenterally

Explanation -Opioids



- ❧ Moderate-to-severe pain is treated with opioids, with or without NSAIDs and adjuvant medications.
- ❧ Codeine- equivalent opioids, such as oxycodone and hydrocodone, are used for moderate pain.
- ❧ When opioids are given for the first time for severe pain, morphine sulfate or hydromorphone should be used.
- ❧ Other morphine-equivalent opioids include oxymorphone, levorphanol, meperidine, fentanyl, and methadone.

Explanation - Meperidine



- ❧ Parenteral meperidine (Demerol) should no longer be used as first-line treatment of acute pain in SCD because of CNS toxicity related to its metabolite, normeperidine
- ❧ Because normeperidine is excreted by the kidneys, meperidine is contraindicated for patients with impaired renal function and those on monoamine oxidase inhibitor antidepressants.

Arrival at emergency department

Assess the pain

Severe acute pain

Assess for common acute pain states associated with SCD

Determine type of pain (onset, duration, frequency)

Typical pain?

No

Determine related symptoms (look for infections, complications, other comorbidities, and precipitating factors)

Determine cause

Related to SCD?

No

Assess pain, treat, and conduct complete workup to determine etiology

Yes

Determine pain characteristics (intensity, location, and quality based on self-report)

Obtain treatment history: home meds, acute pain, hospital Rx, meds past 24 hrs, out-of-home meds?

Examine pertinent physical factors

Summarize assessment profile and select treatment (based on characteristics of episode, prior treatment history, and physical findings)

Currently on chronic opioid therapy?

No

Start IV loading dose of short-acting opioid

- Adults/children < 50 kg of body weight
- Morphine 0.1-0.15 mg/kg
 - Hydromorphone 0.015-0.020 mg/kg
- Adults/children > 50 kg of body weight
- Morphine 5-10 mg
 - Hydromorphone 1.5 mg

Select medication and loading dose based on overall assessment and prior treatment history.

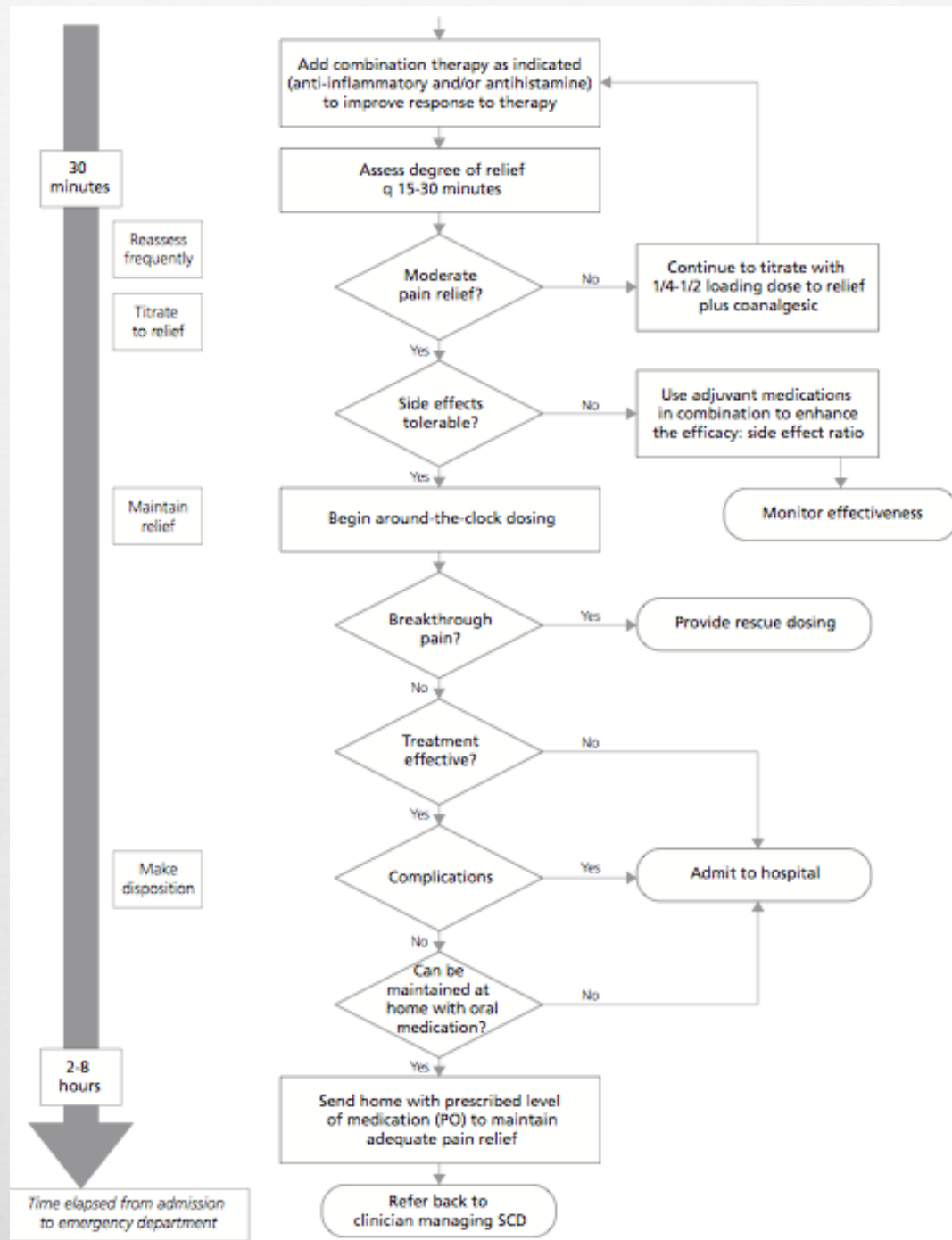
Note: Patient/family often know what medication and dosage have been effective in the past.

Administer by IV (if sufficient venous access) or subcutaneous route (if insufficient venous access)

Time elapsed from admission to emergency department

15-20 minutes

(continued)



References



- ❧ www.uptodate.com - Vasooclusion in Sickle Cell Disease
- ❧ Johnson L, Carmona-Bayonas A, Tick L. Management of pain due to sickle cell disease. *J Pain Palliat Care Pharmacother.* 2008;22(1):51-4
- ❧ The Management of Sickle Cell Disease – NIH PUBLICATION NO. 02-2117 National Heart Lung and Blood Institute June '02
- ❧ Medscape Pediatrics – Sickle Cell Anemia