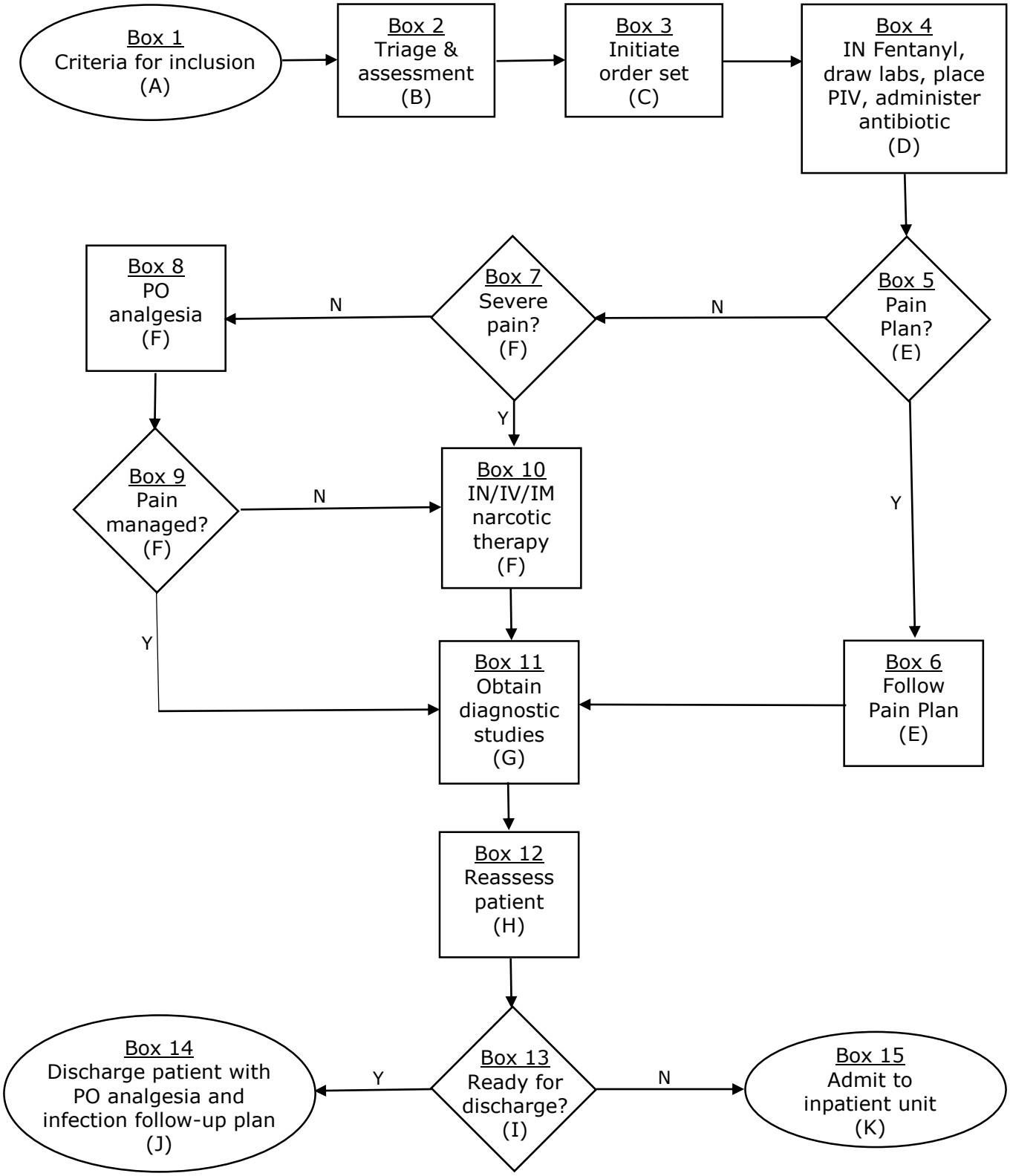
DAYTON CHILDREN'S HOSPITAL

CLINICAL PRACTICE GUIDELINES

DISCLAIMER: This Clinical Practice Guideline (CPG) generally describes a recommended course of treatment for patients with the identified health needs. This CPG is not presented and should not be used as a substitute for the advice of a licensed independent practitioner, as individual patients may require different treatments from those specified, and guidelines cannot address the unique needs of each patient. Dayton Children's shall not be liable for direct, indirect, special, incidental or consequential damages related to the use of this CPG.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(B)
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(C)
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(D)
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(F)
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(F)
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(F)
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(E)
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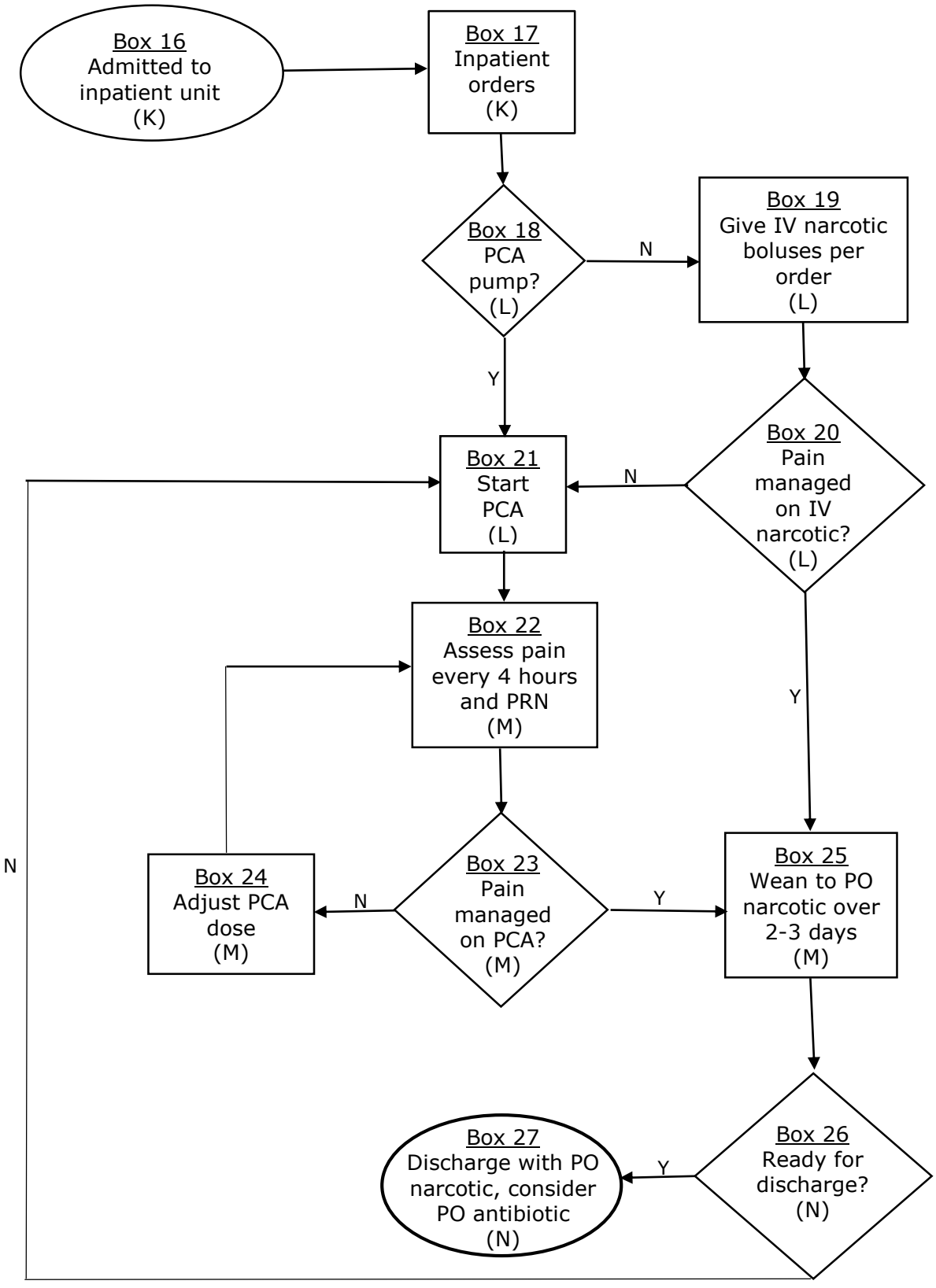
(H)
Box 13
Ready for discharge?
(I)
Box 14
Discharge patient with PO analgesia and infection follow-up plan
(J)
Box 15
Admit to inpatient unit
(K)
N
N
Y
N
Y
Y
Y
N


Box 16
Admitted to inpatient unit
(K)
Box 17
Inpatient orders
(K)
Box 18
PCA pump?
(L)
N
Box 19
Give IV narcotic boluses per order
(L)
Y
Box 20
Pain managed on IV narcotic?
(L)
N
Box 21
Start PCA
(L)
Box 22
Assess pain every 4 hours and PRN
(M)
Y
Box 23
Pain managed on PCA?
(M)
N
Box 24
Adjust PCA dose
(M)
N
Y
Box 25
Wean to PO narcotic over 2-3 days
(M)
Box 26
Ready for discharge?
(N)
Y
Box 27
Discharge with PO narcotic, consider PO antibiotic
(N)

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

A. Box 1: Criteria for inclusion

- Patients with genotype HbSS, HbSC, HbS-beta-zero-thalassemia, HbS-beta-plus-thalassemia, HbSD, and all unknown phenotypes who present with sickle cell pain AND temperature greater than 38.3˚ C (101˚ F)
    - All sickle cell patients with temperature greater than 38.3° C are included, even if a source of fever (e.g. otitis media) has been identified that is typically treated with oral antibiotics in other individuals.
- Patients with HbAS trait, HbAC trait, and HbCC are not routinely followed by a Hematologist and do not have a Pain Management Plan. Additionally, they can be treated as immune-competent individuals. Thus, these patients are excluded from this guideline.

B. Box 2: Triage and assessment by attending physician

- Identification of sickle cell patient with pain and fever should result in immediate triage of the patient so that assessment can be completed within 30 minutes of arrival and subsequent pain medication and empiric antibiotics can be delivered in a timely fashion (within 60 minutes maximum).
- Assessment by attending physician
    - Brief history and physical examination, including vital signs and oxygen saturation
    - During assessment, determine the character, location, and severity of pain and the treatment received (type, dose, and frequency) over the prior 12 hours. Etiologies for pain besides vaso-occlusive crisis should be considered in the evaluation.
    - Fever history
    - Other complications of sickle cell disease should also be addressed, including acute chest syndrome, stroke, sequestration and aplastic crises, and priapism.
        - Acute chest syndrome should be suspected if the oxygen saturation drops greater than 3% from the patient's baseline value.

C. Box 3: Contact hematologist on call to initiate order set, unless the ED attending physician is immediately available to assess the patient and initiate orders.

D. Box 4: Give IN fentanyl, obtain labs, place PIV, and administer antibiotic

- Administer initial dose of IN fentanyl before drawing labs and attempting IV access

- Initial dose: 1.5-2 mcg/kg, maximum dose of 100 mcg

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

D. Box 4 (cont.):

- If unable to obtain IV access after 30 minutes, contact attending physician for subsequent dose of IN fentanyl and continue to attempt IV placement
- Subsequent doses: 0.5-1 mcg/kg every 15 minutes as needed

- Obtain blood culture, CBC with differential, and reticulocyte count and place PIV. Patients in severe pain who require IV medication and/or admission should also have a BMP and type and screen drawn.
  - If blood culture is not obtained within 45 minutes of presentation, administer antibiotic first, and then continue to pursue blood culture.
- Ceftriaxone (Rocephin) 50-100 mg/kg IV should be administered within 60 minutes of presentation to patients who have no known contraindication to this drug.
  - Ceftriaxone (Rocephin) can be given IM if PIV placement is not successful within 45 minutes of arrival in the ED. (Note that the injection may need to be divided into multiple syringes if the maximum volume for an IM injection is exceeded based on patient's weight.)
  - If patient has a contraindication to ceftriaxone, then give ampicillin/sulbactam (Unasyn) 50 mg/kg IV.
  - If patient has a contraindication to ceftriaxone (Rocephin) and PIV placement is not successful within 45 minutes of arrival in the ED, consider ciprofloxacin PO (20 mg/kg).
  - If patient has a contraindication to penicillins and cephalosporins, consult the ED attending physician and consider ciprofloxacin IV (10 mg/kg) or PO (20 mg/kg).

E. Box 5 and 6: Every current patient of the West Central Ohio Comprehensive Sickle Cell Center at Dayton Children's Hospital should have an updated Pain Management Plan in EPIC (under the Letters tab).

a. The Pain Plan is updated at the patient's annual comprehensive visit and on an as-needed basis, based on input from the patients and families.
b. If the patient's weight has changed dramatically from the weight on the Pain Plan, medication dosages may require adjustment.

- If the Pain Plan is outdated (greater than one year since revision) or missing, then proceed to Box 7.

F. Box 7, 8, 9, and 10: Determine the pain severity through clinical assessment and patient self-report

- Mild to moderate pain in clinic
    - Administer PO analgesia based on recommendations on the patient's Pain Management Plan.

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

F. Box 7, 8, 9, and 10 (cont.):

- Reassess pain 45-60 minutes after administration of PO analgesia. If pain is managed, proceed to Box 11. If pain is not managed on PO analgesia, proceed to Box 10.
- Encourage liberal oral hydration

- Severe pain in clinic or emergency department
    - Administer IV narcotics. If unable to obtain IV access, pain medication may be given IM.
    - Morphine sulfate is drug of choice, unless specifically contraindicated
        - Initial dose: 0.10-0.15 mg/kg, to maximum dose of 10 mg
        - Subsequent doses: half of initial dose (0.05-0.075 mg/kg) every 15-30 minutes as needed. Subsequent dose amounts are determined from the degree of pain relief and the amount of sedation produced by the initial dose.
    - May give fentanyl or hydromorphone (Dilaudid) instead of morphine sulfate if patient has better pain control on those narcotics. Do not give meperedine (Demerol).
    - Acute chest syndrome should be suspected with sternal chest pain accompanied by lower respiratory tract symptoms, decreased O_2 saturation, tachypnea, chest pain, cough, or dyspnea. If acute chest syndrome is suspected, order pulse oximetry, cardiorespiratory monitoring, incentive spirometry, and oral azithromycin. Blood transfusion may be indicated based on patient's clinical presentation.
- Reassess for pain, respiratory depression, and excessive sedation 15-30 minutes after administration of IV narcotics.
    - Respiratory depression is defined as a decrease of 4 or more percent oxygen saturation from the baseline value.
    - Sleeping while maintaining oxygenation does not indicate excessive sedation.
    - Provide IV hydration
        - Hydrate at 1 to 1.5 maintenance using low-sodium fluids (i.e., ¼ NS).
        - Be cautious if acute chest syndrome is suspected, as over-hydration may precipitate pulmonary edema.

G. Box 11: Obtain other diagnostic studies as appropriate or relevant, based on the patient's symptoms

- Radiology: chest x-ray (for acute chest syndrome/pneumonia) and other imaging studies as indicated
- Diagnostic lumbar puncture for patients with meningeal signs, neurologic deficit, and/or altered sensorium

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

G. Box 11 (cont.):

- Throat culture
- Stool culture
- Urine culture and urinalysis
- Other

H. Box 12: Reassess patient

- The goal is not for the patient to be pain-free. Rather, the pain should be well-managed at or near the patient's pain baseline, with the expected ability to manage at home with PO medications.
- If pain is well-managed after three IV/IM doses of narcotics, confer with the family and transition patient to PO medications. Consider administering initial dose of oral narcotic analgesia (Tylenol #3 or Norco) before patient leaves the hospital.
- If pain is not well-managed after three IV/IM doses of narcotics, **confer with the hematologist on call** and proceed to Box 13.
- Review patient's laboratory results, diagnostic studies (if applicable), and current clinical status.

I. Box 13, 14, and 15: Ready for discharge?

- Patient may be discharged from the ED if <u>all</u> of the following criteria are met:
  - Family is comfortable with the discharge plan
  - Age greater than 12 months
  - Nontoxic, well-appearing
  - Patient is at or near pain baseline
  - Stable vital signs with O_2 sats $\geq$ 92% on room air 60 minutes after antibiotic administration is complete
  - Tolerating oral intake
  - No concern for complications (i.e., splenic sequestration, acute chest syndrome, or pain crisis requiring IV analgesia)
  - Hgb > 5 with no drop greater than 2 g
  - WBC > 5,000 and < 30,000
  - Chest x-ray (if applicable) without infiltrate

- Patient has reliable follow-up in place (i.e., working telephone, available transportation, no history of missed appointments or noncompliance with medications)

- If the above discharge criteria are not met, the patient should be admitted to the Hem/Onc unit.

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

J. Box 14: Patients who meet the discharge criteria should be given a follow-up plan

- For patient with no known source of infection, the discharge physician should contact the hematologist on-call and have the patient follow-up in 24 hours in the Hematology Clinic. (If follow-up is to occur on a day when the Clinic is closed, patient will follow-up in the Almost Home Unit.) **Leave PIV in place**, if possible, for a second dose of ceftriaxone.
- For patient with bacterial source of infection (i.e., Streptococcus, otitis media, urinary tract infection, impetigo), the discharge physician should treat the infection with the appropriate oral antibiotic and have the patient follow-up in the Hematology Clinic after the course of antibiotics is complete.
- For patient with viral source of infection, the discharge physician should contact the hematologist on-call and have the patient follow-up in 24 hours in the Hematology Clinic. (If follow-up is to occur on a day when the Clinic is closed, patient will follow-up in the ED.) **Leave PIV in place**, if possible, for a second dose of ceftriaxone.
- All patients should call their physician or go to the ED for pain, shortness of breath, lethargy, dizziness with standing, or for worsening symptoms.

K. Box 15, 16, and 17: Patients who do not meet the discharge criteria should be admitted to the inpatient unit.

- Inpatient orders should include:
  - Daily CBCs and a daily blood culture for persistent fever
  - Vital signs and oxygen saturation every 2 hours x 2, then every 4 hours
  - Cardiorespiratory monitoring
  - A daily stool softener
  - A nonsteroidal anti-inflammatory medication
  - Incentive spirometry every hour during waking hours
  - Ambulation three times per day
  - PRN doses of antiemetics and antihistamines should also be ordered for narcotic-induced nausea and itching
  - Ceftriaxone (or ampicillin/sulbactam) should be continued daily until cultures are negative for a minimum of 48 hours.

L. Box 18, 19, 20, and 21: Is a PCA pump available and appropriate for the patient?

- If a PCA pump is available and appropriate, start PCA per orders
- If a PCA pump is not available and/or appropriate, give IV narcotic boluses per orders
    - If pain is well-managed on IV narcotic boluses, wean the patient to PO analgesia over 2-3 days and proceed to Box 23

SICKLE CELL PAIN WITH FEVER CLINICAL PRACTICE GUIDELINE NOTES

L. Box 18, 19, 20, and 21 (cont.):

- If pain is not well-managed on IV narcotic boluses, start the patient on a PCA pump per orders

M. Box 22, 23, 24, and 25: Assess patient's pain every four hours and PRN while on the PCA pump.

- If the pain is not well-managed on the PCA pump, adjust the PCA dose and continue pain assessments every four hours and PRN. Alternative pain management may also be considered, including epidural analgesia and intrathecal morphine. May refer to a pain specialist.
- If the pain is well-managed on the PCA pump, wean the patient to PO analgesia over 2-3 days and proceed to Box 23.

N. Box 26 and 27: Is patient ready for discharge?

- Patient may be discharged from the Hem/Onc unit if <u>all</u> of the following criteria are met:
    - Comfortable, well-appearing
    - At or near pain baseline
    - Stable vital signs with O_2 sats $\geq$ 92% on room air 60 minutes after last dose of medication is administered
    - No concern for complications (i.e., splenic sequestration, acute chest syndrome)
    - Chest x-ray (if applicable) without infiltrate
- Arrange follow-up appointment and consider oral antibiotic.
- Discharge patient with sufficient oral pain medication for next 5-7 days (consider combination of an oral narcotic and NSAID, i.e., Tylenol #3 and ibuprofen or Naprosyn).
- If the above discharge criteria are not met, proceed to Box 21.

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