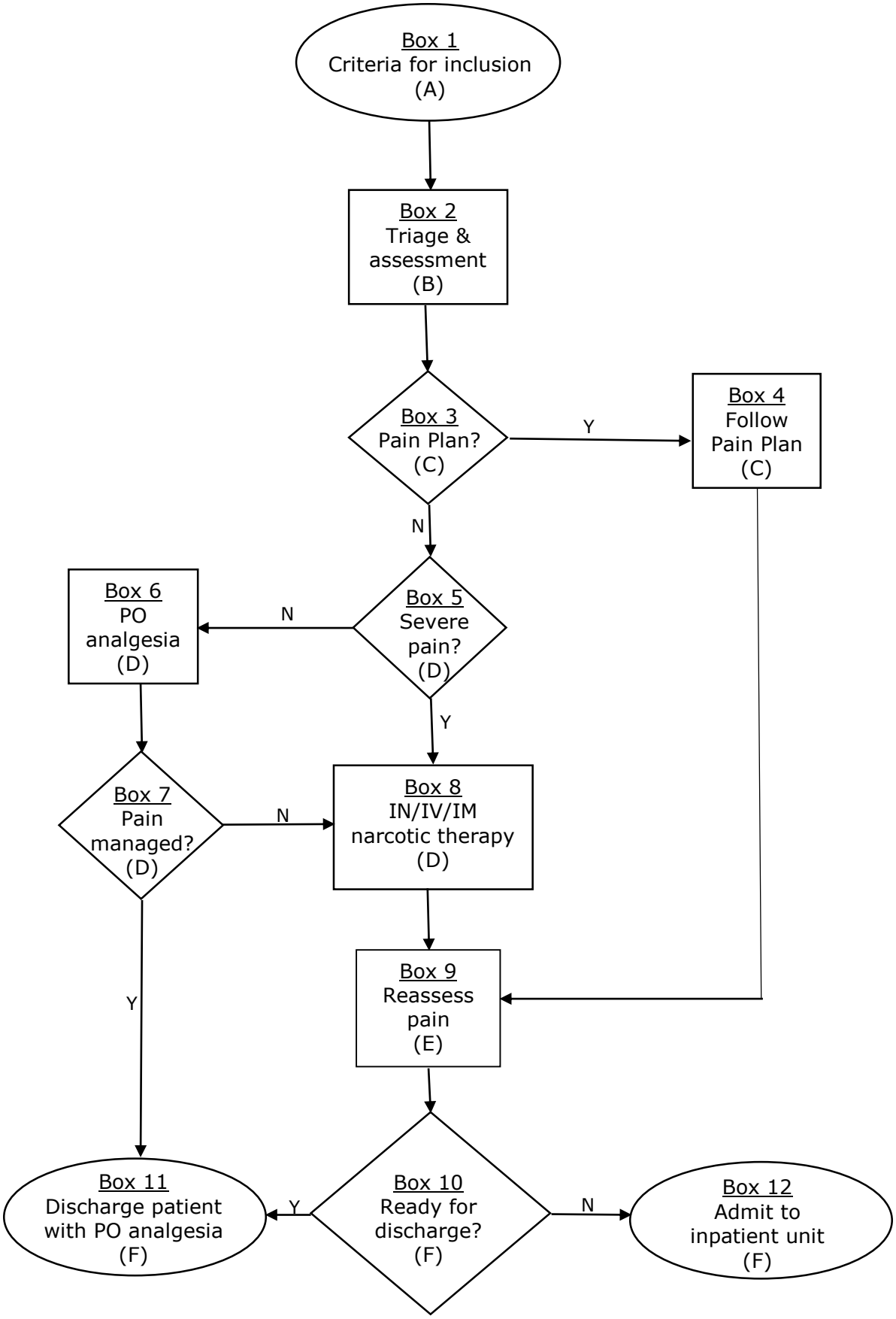
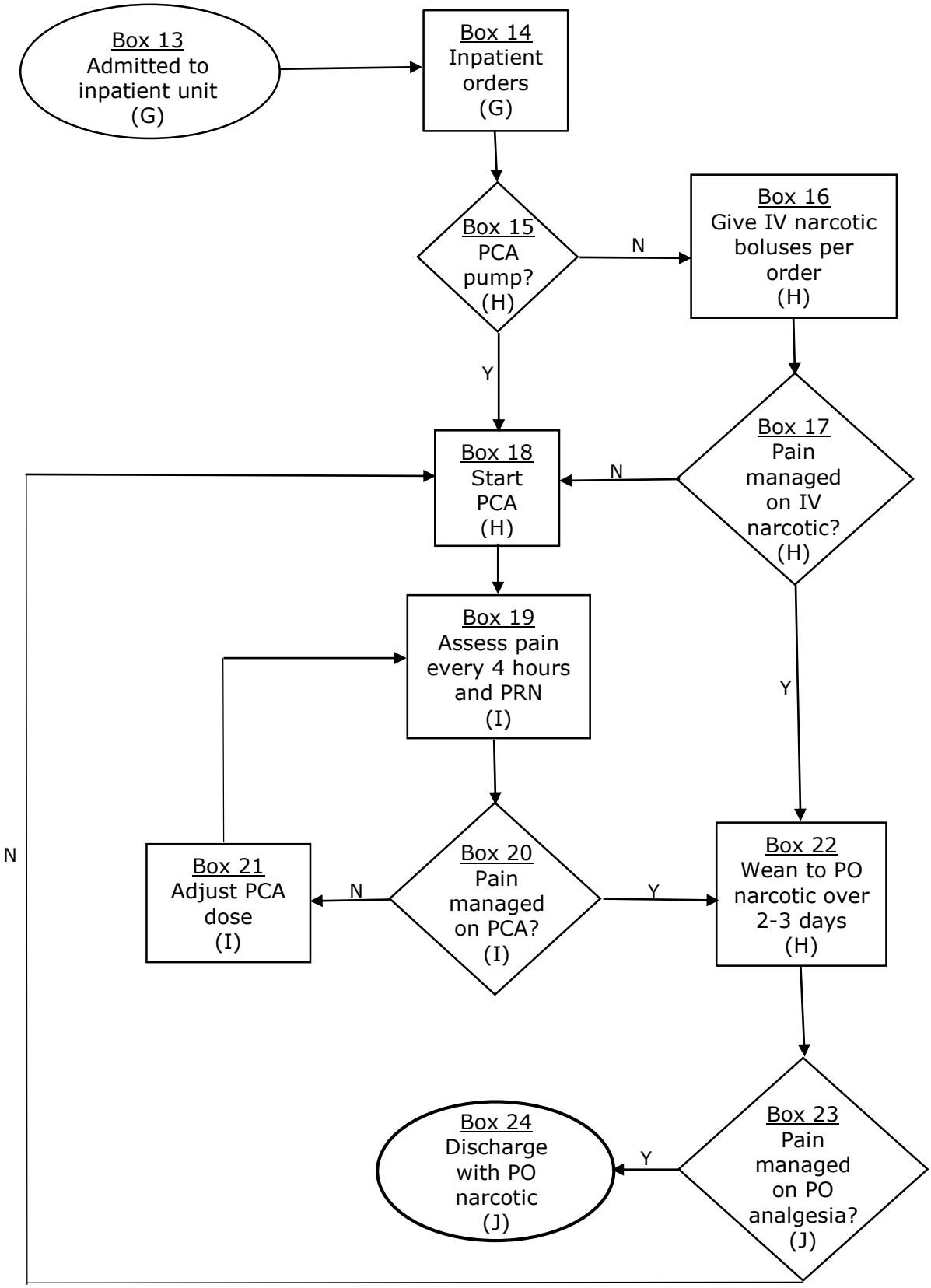
DAYTON CHILDREN'S HOSPITAL

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.


Box 1
Criteria for inclusion
(A)
Box 2
Triage &
assessment
(B)
Box 3
Pain Plan?
(C)
Y
Box 4
Follow
Pain Plan
(C)
N
Box 5
Severe
pain?
(D)
N
Box 6
PO
analgesia
(D)
Y
Box 7
Pain
managed?
(D)
N
Box 8
IN/IV/IM
narcotic therapy
(D)
Box 9
Reassess
pain
(E)
Y
Box 10
Ready for
discharge?
(F)
Y
Box 11
Discharge patient
with PO analgesia
(F)
N
Box 12
Admit to
inpatient unit
(F)


Box 13
Admitted to inpatient unit
(G)
Box 14
Inpatient orders
(G)
Box 15
PCA pump?
(H)
N
Box 16
Give IV narcotic boluses per order
(H)
Y
Box 17
Pain managed on IV narcotic?
(H)
N
Box 18
Start PCA
(H)
Box 19
Assess pain every 4 hours and PRN
(I)
Y
Box 20
Pain managed on PCA?
(I)
N
Box 21
Adjust PCA dose
(I)
N
Y
Box 22
Wean to PO narcotic over 2-3 days
(H)
Box 23
Pain managed on PO analgesia?
(J)
Y
Box 24
Discharge with PO narcotic
(J)

SICKLE CELL PAIN 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 are covered by this guideline.
- Patients with HbAS trait, HbAC trait, and HbCC are not routinely followed by a Hematologist and do not have a Pain Management Plan. Thus, these patients are excluded from this guideline.

B. Box 2: Triage and assessment by attending physician

- Identification of sickle cell patient with pain should result in immediate triage of the patient so that assessment can be completed within 30 minutes of arrival and pain medication can be delivered in a timely fashion (within 60 minutes of the pain assessment). Fever should be evaluated and treated according to the **Sickle Cell Pain with Fever Clinical Practice Guideline**.
- Assessment by attending physician
    - 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.
    - Other complications of sickle cell disease should also be addressed, including fever, acute chest syndrome, stroke, sequestration and aplastic crises, and priapism.
    - Vital signs and oxygen saturation should be obtained on all patients.
        - Acute chest syndrome should be suspected if the oxygen saturation drops greater than 3% from the patient's baseline value.
    - Labs are optional, at the discretion of the attending physician. However, those in severe pain who require IV medication and/or admission should have a CBC with differential, reticulocyte count, BMP, and type and screen drawn.

C. Box 3 and 4: 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).

- 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.
- If the patient's weight has changed dramatically from the weight on the Pain Plan, medication dosages may require adjustment.

SICKLE CELL PAIN CLINICAL PRACTICE GUIDELINE NOTES

C. Box 3 and 4 (cont.):

- If the Pain Plan is outdated (greater than one year since revision) or missing, then proceed to Box 5.

D. Box 5, 6, 7, and 8: 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.
    - 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 8.
    - Encourage liberal oral hydration
- Severe pain in clinic or emergency department
    - Administer initial dose of IN fentanyl before attempting IV access
        - Initial dose: 1.5-2 mcg/kg, maximum dose of 100 mcg
        - 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
    - 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.

SICKLE CELL PAIN CLINICAL PRACTICE GUIDELINE NOTES

D. Box 5, 6, 7, and 8 (cont.):

- 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.

E. Box 9: Reassess pain

- 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 12.

F. Box 10, 11, and 12: 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
    - Patient is comfortable, well-appearing
    - Patient is at or near pain baseline
    - Patient has 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
- If the above discharge criteria are not met, the patient should be admitted to the Hem/Onc unit.

G. Box 13 and 14: Inpatient orders should include:

- Vital signs and oxygen saturation every 2 hours x 2, then every 4 hours

SICKLE CELL PAIN CLINICAL PRACTICE GUIDELINE NOTES

G. Box 13 and 14 (cont.):

- 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

H. Box 15, 16, 17, 18, and 22: 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
    - If pain is not well-managed on IV narcotic boluses, start the patient on a PCA pump per orders

I. Box 19, 20, and 21: 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.

J. Box 23 and 24: Is pain managed on PO analgesia?

- 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 for 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

SICKLE CELL PAIN CLINICAL PRACTICE GUIDELINE NOTES

J. Box 23 and 24 (cont.):

- 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 18.

References

Barclay, L. (2010). Intranasal medication delivery in children. Retrieved from http://www.medscape.com/viewarticle/726524_print

Barrett, M.J., Cronin, J., Murphy, A., et al. (2012). Intranasal fentanyl versus intravenous morphine in the emergency department treatment of severe painful sickle cell crisis in children: Study protocol for a randomized controlled trial. *Trials, 13*, 74-81.

Buchannan, G., Yawn, B., et al. (2014). Evidence-based management of sickle cell disease, expert panel report. Retrieved from http://www.nhlbi.gov/guidelines

Kavanaugh, P., Sprinz, P., Wolfgang, T., et al. (2015). Improving the management of vaso-occlusive episodes in the pediatric emergency department. *Pediatrics, 138*, 1016-1025.

Lovett, P., Sule, H., and Lopez, B. (2014). Sickle cell disease in the emergency department. *Emergency Medicine Clinics of North America, 32*, 629-647.

Mathias, M. and McCavit, T. (2015). Timing of opioid administration as a quality indicator for pain crises in sickle cell disease. *Pediatrics, 135* (3), 475-482.

Murphy, A., O'Sullivan, R., Wakai, A., Grant,T.S., et al. (2014). Intranasal fentanyl for the management of acute pain in children (review). Retrieved from http://onlinelibrary.wiley.com/doi/10.1002/14651858.CD009942.pub2/epdf/standard

Prommer, E. and Thompson, L. (2011). Intranasal fentanyl for pain control: Current status with a focus on patient considerations. *Patient Preference and Adherence, 5*, 157-164.

Tanabe, P., Hafner, J., Martinovich, Z., and Artz, N. (2012). Adult emergency department patients with sickle cell pain crisis: Results from a quality improvement learning collaborative model to improve analgesic management. *Academic Emergency Medicine, 19*, (4), 430-438.