

INPATIENT FEVER MANAGEMENT

- Continue **IV/PO antibiotics** as started in ED
- Fluids** to correct deficit, then IV/PO @ 1.25x maintenance or IV/PO @ 2/3 - 1x maintenance for pts w/ fluid sensitivity, ACS, or resp sx
- O2 only for documented hypoxemia or resp symptoms (keep O2 sats ≥93%)
- Segregate from anemic or immunosuppressed pts & pregnant caretakers if parvovirus is suspected
- CXR for respiratory deterioration or persistent ↓ in O2 sats while awake
- Consider CBC/diff & retic 24 hrs after admission or with any deterioration
- Blood culture if toxic or temp spike after being afebrile x 24hr
- Ambulation at least 2x per shift unless acute splenic sequestration
- Incentive spirometry** 10x/hour while awake
- Consider BiPAP overnight & when in bed as tolerated if pulmonary infiltrate, resp sx, chest wall pain, or history of ACS
- Plan discharge after 2nd dose of ceftriaxone if pt is stable & has adequate follow-up.
Do not give Oral antibiotics upon discharge unless treatment for focal infection requiring longer therapy (ie. pneumonia, bacteremia, osteomyelitis).
- Restart prophylactic antibiotics on discharge, if applicable

BLOOD TRANSFUSIONS

STRAIGHT TRANSFUSIONS

Purpose: ↑ RBC mass & O2 carrying capacity, ↓ proportion of sickle cells (dilution). Pt receives transfused blood only. None of their own blood is removed. Used correctly, can prevent organ damage and save life of SCD pts.

Type of blood: Phenotypically matched, (ABO, Rh-D, Kell, C, E), sickle negative, leukodepleted irradiated packed cells are blood product of choice. More extensive phenotyping for pts on chronic transfusion.

Indications:

- Acute anemia: Hct < 17 or Hct + retic < 30; Hb ↓ >2 g/dl from baseline without a compensated reticulocytosis
- Acute chest syndrome (ACS): To prevent progression of ACS to acute resp failure. Single lobe involvement w/ resp sx in pts w/o history of ACS. Evaluate in asymptomatic pts w/history of ACS and new infiltrate
- Hypoxemia (PaO2 < 70 mmHg on room air) or ↓ >10% from baseline
- Symptomatic anemia regardless of Hct (heart failure, ↑HR, dyspnea, marked fatigue)
- Aplastic crisis
- Pre-op with general anesthesia to ↑ Hct to 28-35%. Pts w/ hx of severe ACS or lung disease may be exchanged
- Splenic sequestration
- Prolonged and complicated vaso-occlusive episodes

Amount of blood: Goal = ↑ Hct to 30% (target range 28-33%). Higher values (>35%) cause hyperviscosity and ↑ risk for stroke in SCD pts

Always check your calculations with someone else!

Rule of thumb: 3 ml/kg of PRBC will ↑ Hb by 1 g/dl and Hct by 3%;
10 ml/kg of PRBC will ↑ Hb by 3 g/dl and Hct by 10%

Vol of PRBC (ml) = EBV (80ml) x WT (Kg) x Desired Hct – Actual Hct
Hct of PRBC (60%)
EBV: estimated blood volume

Example: 20 kg child with Hct of 15%: PRBC (ml) = 80 x (30% – 15%) x 20 kg = 400ml
60%

How to order: 1 unit PRBC contains ~250-350 ml and has Hct ~50-60%. Order blood as # units (adults) or # mLs (pediatrics).

Rate of transfusion

- history of fluid intolerance or CHF 1-2 ml/kg/hour
- no history of fluid intolerance or CHF 3 ml/kg/hour
- on chronic transfusion, not otherwise ill 4-5 ml/kg/hour

Rapid replacement is required for acute blood loss or shock.

Rule of thumb in severe compensated anemia:
Give “X” mL/kg aliquot, where X = Hb. (If Hb = 5, transfuse 5 ml/kg over 4 hrs)

EXCHANGE TRANSFUSION

Purpose: REMOVE SICKLE CELLS TO 30-40% & replace w/normal RBCs w/out ↑ whole blood viscosity or chronic iron burden. **Manual exchange** (see Transfusion Guidelines for Procedure) vs. **Automated erythrocytapheresis** automated exchange device (coordinate w/blood bank, only week days)

Indications: ACS (symptomatically decompensated, multiple lobe infiltrate); Suspected or presenting w/ acute stroke or TIA; Pre-op w/general anesthesia + hx of severe ACS or pulmonary disease; Chronic transfusion protocol in pts w/elevated ferritin; Conditions unresponsive to simple transfusion (severe VOC, priapism, splenic sequestration)

Sickle Cell Disease Summary Card 2010

Pediatric Hematology

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INPATIENT ORDERS FOR ACUTE PAIN EPISODE

For pain management see: **Inpatient Management of Acute Painful Episodes** algorithm (next panel on this sheet)

See Pediatric PCA Guideline on BMC intranet under Pharmacy for more info. Additional information can be found under Pediatric Hematology CPG for pain management on the New England Pediatric Sickle Cell Consortium website (NEPSCC.org)

PCA DOSING	MORPHINE	HYDROMORPHONE
Basal Rate (mg/kg/hr)	0.02 – 0.04	0.003 – 0.007
PCA dose (mg/kg)	0.015	0.0025
Lockout period	6 min	6 min
Bolus Dose (mg/kg)	0.05	0.008

Adjust PCA Q 30-60 min to capture the pain.

Monitoring

- Nurse to evaluate pt Q4hrs (while awake) and document pain intensity on bedside flow sheet.
- Symptomatic relief: apply warm packs to painful areas
- Stool softener +/- laxative while on narcotics
- Continue with routine prescribed home medications
 - Hydroxyurea if taking
 - Folic acid if taking
 - Home antibiotics: - if < 2 year PCN 125 mg PO BID
- if > 2 years PCN 250mg PO BID
 - PCN is d/c at 5 y/o, except in pts w/splenectomy
 - D/C home antibiotics if on hospital antibiotics
- Antibiotics as per fever management guidelines

Respiratory

- Incentive spirometry “every TV commercial” (10x per hour)
- Early ambulation at least 2x per shift, days and evenings
- Oxygen to keep sats ≥ 93%, Consider BiPAP if history of ACS or chest wall pain. Remember: everyone drops their sats while sleeping. All patients to sleep with the back of their bed elevated
- Albuterol nebs TID for pts w/history of asthma or acute resp sx
- CXR for fever associated w/resp distress, desats, or abnormal lung exam

Fluids

- IV+PO @ 1.25x maintenance. IV+PO @ 2/3 – 1x maintenance if history of fluid sensitivity, ACS, or presenting w/resp sx
- Strict I/O's and daily weight

Labs

- CBC/diff, retic count every 48 hrs if clinically stable
- Blood cultures with CBC/diff for first temp > 101.5°F, then every 24hr with temp spike ≥101.5°F (38.5°C)

In the event of severe somnolence or low RR <12: Stimulate, apply O2, disconnect PCA.

Notify pedi & heme attendings.

Naloxone: dilute 0.4 mg in 10 ml NS. Give to effect.

If patient experiences any side effects (pruritus, N/V, constipation) from narcotic infusion consider

Naloxone drip: 0.001 mg/kg/hour

Patient will require a second IV for a Naloxone infusion.

(Pharmacy standard concentration = 0.04 mg/ml) (2mg in 50ml NS)

INPATIENT MANAGEMENT OF ACUTE PAINFUL EPISODE

All patients being admitted for acute painful episode should receive CONTINUOUS IV OPIOID. PRN MEDICATION IS NOT ACCEPTABLE. Start PCA with morphine if no contraindication.

Upon arrival to the floor, if the pt did not come with a continuous opioid infusion PCA, or > 1 hour has elapsed since last opioid dose, use patient specific dosing (if available):
give morphine: 0.05 mg/kg IV (max 5 mg) OR hydromorphone: 0.008 mg/kg IV (max 2 mg)

Continuous Morphine

OR

Hydromorphone

AND

Ketorolac

AGE < 7 years

Continuous IV opioid + PRN bolus (as a separate order)

Basal rate: 0.02 – 0.04 mg/kg/hr

AGE ≥ 7 years

PCA and PRN bolus.

Basal rate: 0.02-0.04 mg/kg/hr

PCA dose: 0.015 mg/kg/dose Q 6 min

Hourly max = Basal rate + 0.15 mg/kg (10 PCA doses) = 0.19 mg/kg/hr

Hydromorphone dosing

(approximately 1/6th of morphine dose)

Basal rate: 0.003 – 0.007 mg/kg/hr

PCA dose: 0.0025 mg/kg/dose Q6min

Hourly max = Basal + 0.025 mg/kg (10 PCA doses) = 0.032 mg/kg/hr

Use only if opioid treatment fails to provide adequate relief

0.5 mg/kg/dose IV Q6h for 48 hrs (Max 30 mg/dose)

When discontinued, consider ibuprofen.

RE-EVALUATE EVERY 30 MIN UNTIL PAIN UNDER CONTROL

Persistent pain

1st evaluation
Pushing PCA >3 x hr, **give morphine 0.05 mg/kg bolus**

2nd evaluation
Give morphine 0.03 mg/kg bolus & then ↑ basal rate by 20%

hydromorphone 0.008 mg/kg

hydromorphone 0.005 mg/kg

Well controlled pain

Continue current management and re-evaluate in 4-6 hours

RE-EVALUATE EVERY 4-6 HOURS AND EVERY 1-2 HOURS AFTER EACH CHANGE

Persistent pain

Morphine 0.03 mg/kg bolus & then ↑ basal rate by 10-20% every 4-8 hours as tolerated

hydromorphone 0.005 mg/kg

Basal rate should be > 2/3 of Total Opioid Dose

Calculate total dose for previous 6 hours
Total Opioid Dose = Total bolus (PCA + bolus) + Total Basal

- Ideally Total Bolus < 1/3 of Total Opioid
- Nighttime basal may be at a higher rate than daytime basal to facilitate comfort during sleep

Transition to oral analgesia:

- Continue PCA/bolus dosing as most recently prescribed.
- Calculate previous 24hr intake of basal morphine, divide by 3 for Oral equivalent dose. Provide that amount Q8h as oral long-acting morphine. (example: 2mg/hr IV x 24hr = 48mg + 3 = 15mg PO Q8h)
- When pain is controlled by this schedule, change PCA/bolus dosing to oxycodone scheduled Q4hr

Well controlled pain

Is total **morphine** < 0.025 mg/kg/hr?

or **hydromorphone** < 0.004 mg/kg/hr?

YES

NO

↓ basal rate by 10-20% every 4-8 hr as tolerated

Decisions regarding care of any pt should be made with respect to the pt’s individual presenting circumstances. Specific medications & dosing must always be reviewed carefully considering hx of allergy or adverse reactions.

EMERGENCY DEPARTMENT FEVER MANAGEMENT

This section applies to pts >3 months with Temp ≥ 101.5°F

The febrile patient with sickle cell disease (SCD) should be evaluated and treated as a level II patient.

DO NOT DELAY ANTIBIOTICS, GIVE IMMEDIATELY AFTER OBTAINING BLOOD CULTURES, NO LONGER THAN 1 HOUR AFTER PRESENTATION TO ED/CLINIC

INITIAL EVALUATION
Rapid triage: PLACE IMMEDIATELY IN EXAM ROOM AND EVALUATE:

- History and physical exam
- CBC w/diff & retic count, blood cultures
- UA and cx (catheterize males < 6 months, females < 2 years), throat culture, stool specimens, viral panel, other studies as indicated
- CXR (PA and lateral) in pts with respiratory symptoms or hypoxia (including O2 sats > 3% below baseline)
- Clot drawn and held by blood bank

GIVE ANTIBIOTICS (PRIOR TO SENDING TO CXR)

Do not delay antibiotics while awaiting lab results and CXR
• Ceftriaxone 50 mg/kg IV or IM (if no IV access), max 1gm Q24h; Use meningitic dosing (100 mg/kg/day ÷ Q12h, max 2gm Q12h) if CNS infection is suspected

If allergic to ceftriaxone or if on other 3rd generation cephalosporin, pt needs to be admitted:

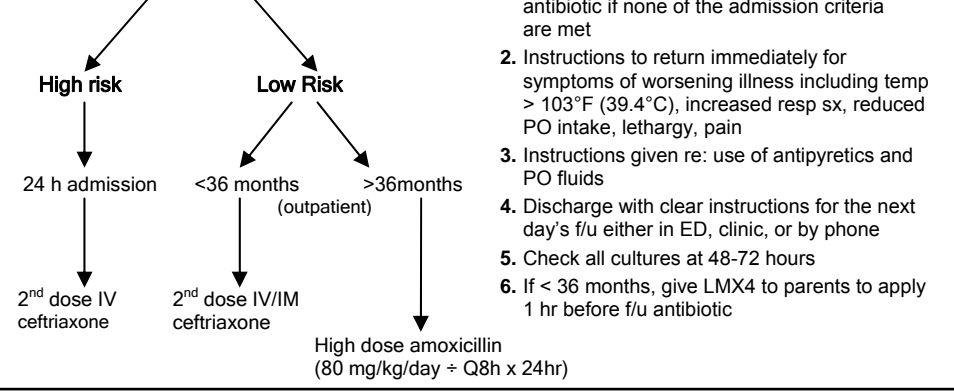
- Azithromycin 12 mg/kg/dose IV or PO Q24h
- Clindamycin 10-15 mg/kg/dose IV Q8h (max: 600 mg/dose)
- Vancomycin 15-20 mg/kg/dose Q8hr for septic shock or meningitis. Consider in pts w/central line/porta-cath and history of infection with resistant organisms. Add Azithromycin PO for pts with positive CXR, or respiratory symptoms

CONSIDER ADMISSION if any of the following high risk criteria are present:

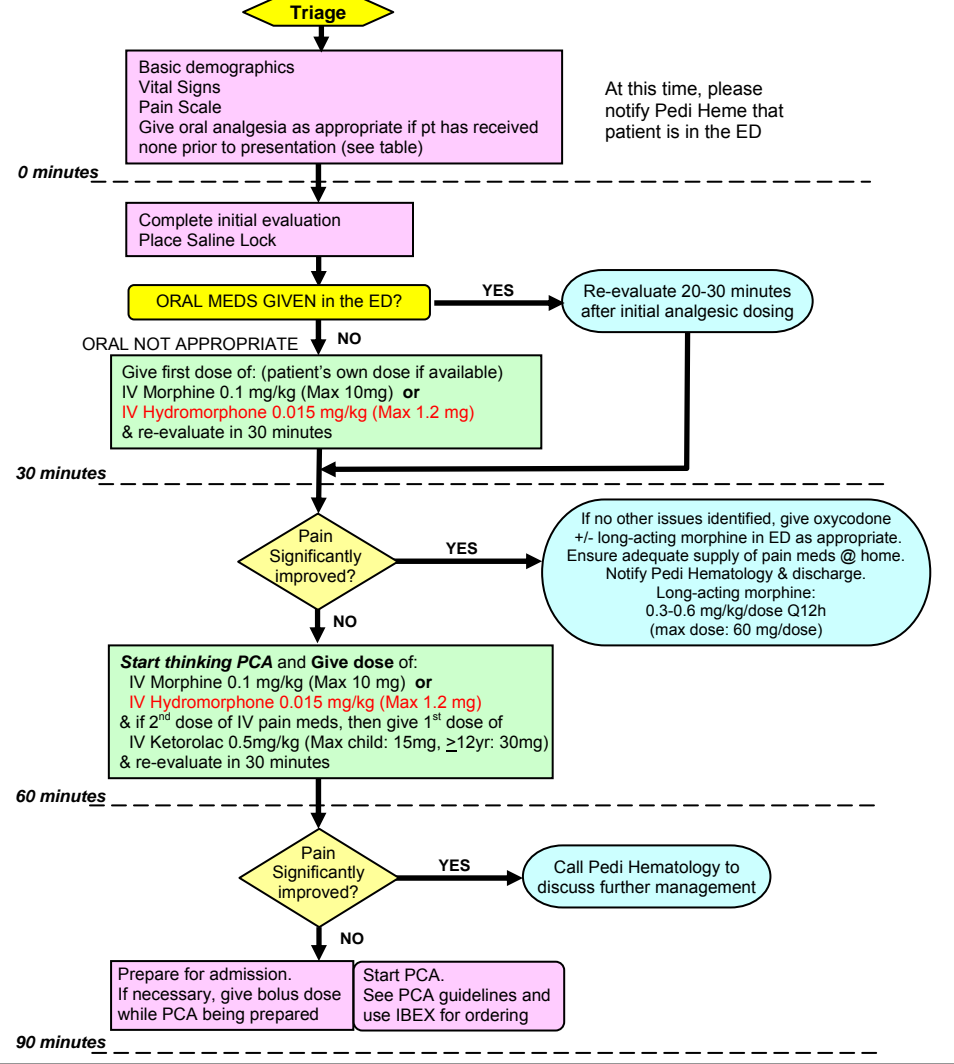
- Age < 12 months
- Constitutional findings: Fever > 104°F (40°C)
 - Toxic appearance
 - Hypotension or poor perfusion
 - Unable to maintain oral hydration
- Respiratory findings: hypoxia (O2 sats > 3% below baseline), new pulmonary infiltrate (acute chest syndrome)
- Hematologic findings: Hct ≤ 18% or ≥ 5 points below baseline; WBC > 30,000 or < 5,000; Platelets < 100,000
- Evidence of another complication: severe VOE, aplastic crisis, splenic sequestration, neurologic findings, priapism
- OTHER: History of prior episode of bacteremia or sepsis
 - Missing or delayed immunizations (PCV, pneumococcal, Hib)
 - Less than 5 y/o and not compliant w/antibiotic prophylaxis
 - History of non-compliance w/medical care or keeping appointments
 - Poor likelihood of outpatient follow up (no phone or transportation)
 - Family is unable to care for patient at home
 - Multiple ED visits for same episode of illness
 - Unable to receive initial 24 hrs of coverage from single dose of medication due to allergies

Be sure to re-evaluate your patients every 1-2 hours after initial evaluation as their clinical status can deteriorate rapidly.

INITIAL ED EVALUATION OF FEVER



EMERGENCY DEPARTMENT PAIN MANAGEMENT



SICKLE CELL DISEASE HISTORY

- See patient-specific data sheet if available from Pediatric Hematology for details
- Minimize history taking to pertinent information relevant to current admission until patient's pain of illness more under control
- HPI should always include status of:
 - Fever
 - Pain
 - Headache
 - Hydration
 - Respiratory symptoms (difficulty breathing, cough)
- IF IT IS POSSIBLE TO OBTAIN THESE FROM THE RECORDS, THEN DO NOT ASK UP FRONT
 - Past sickle cell history:
 - Usual Hb, usual O2 sat
 - Last hospitalization/ED visit
 - Pain: usual sites, home mgmt, previous opioid use
 - Splenic sequestration, splenectomy
 - Transfusion: last transfusion, complications, reactions, indications, simple or exchange, chronic transfusion, iron overload, chelation therapy
 - Other medical history:
 - RAD/asthma, sleep apnea or snoring
 - T&A or other surgical procedures
 - Cholecystectomy or presence of gallstones
 - Porta-cath placement
 - Current medications
 - Allergies or adverse drug reactions
 - Immunizations including PCV7, PCV13, PCV23 (Pneumovax), Meningovax, and influenza

COMMONLY USED ORAL ANALGESICS

Oral Analgesics:
If patient expresses a preference, provide that one. If not, provide from list below:

Medication	ORAL Dose		Max Dose
Morphine (immediate release)	0.2 - 0.5 mg/kg	Q2-4h	60 mg/dose
Morphine (sustained release) (MS Contin®)	0.3 - 0.6 mg/kg	Q8-12h	60 mg/dose
Methadone	0.1 mg/kg	Q8-12h	12 mg/dose
Oxycodone (immediate release)	0.1 - 0.2 mg/kg	Q4-6h	15 mg/dose
Hydromorphone (Dilaudid®)	0.03 - 0.08 mg/kg	Q3-4h	2 mg/dose
Codeine	0.5 - 1 mg/kg	Q4h	60 mg/dose
Ibuprofen	10 mg/kg	Q4-6h	800 mg/dose
Acetaminophen	15 mg/kg	Q4-6h	1000 mg/dose

OTHER ORAL MEDICATIONS WHILE ON NARCOTICS

Docusate: 5 mg/kg/day ÷ 1-4 doses/day (max 400 mg/day)
Senna: 2.5-10 mL/dose or 1-2 tabs PO QHS or BID, max 2 tabs PO BID
Lactulose: 5-20 grams/day ÷ Q6-8h, patient must be taking PO fluids
Milk of Magnesia: 5-10 mL/dose PO BID
Bisacodyl: 5-10 mg/dose PO/PR Q24h
PEG 3350 (Miralax®): 0.5-1.5 g/kg/dose Q24h, max dose 17gm
Diphenhydramine: 1 mg/kg/dose PO/IV Q6hr PRN, max 50 mg/dose
Famotidine: 0.5 mg/kg/day ÷ BID, max dose 40mg BID
Consider Methylphenidate HCl (Ritalin®) 5 mg PO Q24h before breakfast while on narcotics for oversedation and pain