

 CREDIT VALLEY <small>THE CREDIT VALLEY HOSPITAL</small>	CLINICAL PRACTICE GUIDELINE	PROFESSIONAL PRACTICE
TITLE: Management of Sickle Cell Disease in Children		
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Purpose:

To provide direction for the management of children with sickle cell disease who present to the emergency department with complications of their condition. These guidelines are directed at management of these complications and are not intended to replace long-term management by a Paediatric Haematologist.

Inclusion Criteria:

Paediatric patient with sickle cell disease presenting to the emergency department with:

- A. Acute Painful Episodes (Vaso Occlusive Crisis)
- B. Fever (Rule Out Infection)
- C. Acute Chest Syndrome or Pneumonia

All physicians may initiate this guideline by completing the preprinted physician order sheet titled ["Physician's Orders Paediatric Sickle Cell Disease" #70009.](#)

1. General Recommendations for the Management of Paediatric Sickle Cell Disease

Emergency Management Sickle Cell Disease

- Children presenting at the Emergency Department should be classified at a minimum Level 2 (P-CTAS), and should be assessed by a physician within 15 minutes of arrival.
- A paediatric consultation is encouraged following assessment by the Emergency Physician.
- Patient-specific clinical information should be dictated whenever possible to allow staff timely access to relevant history and treatment data.
- Consent to participate in sharing information through HiNet is recommended.

Hospital Admission

- Children requiring inpatient care should be admitted to a Regional Children's Health Centre.

Surgery

- For any child with Sickle Cell Disease all **Elective** surgical procedures should be performed at a Tertiary Care Centre.
- Emergency surgery or procedural sedation and analgesia requires anaesthesia consultation.

History

- Nature, duration, location and severity of pain, how pain compares to previous crises, analgesics already used, associated symptoms and previous successful experience with analgesics.
- Breathing difficulties, fever, previous episodes of ACS or pneumonia

Physical

- vital signs, oxygen saturation, cardiopulmonary status, hydration status, degree of palor, spleen size, neurologic exam, presence of jaundice and localizing signs of infection.

Laboratory investigations

- Ensure that sickle cell diagnosis is entered on laboratory requisitions.

Consultation with a tertiary care center

- The attending paediatrician should consult with a member of a Tertiary Sickle Cell Team or a Haematology Fellow whenever the following conditions are present:

- Presence of acute chest syndrome
- Positive blood culture result
- Clinical condition is deteriorating
- Blood transfusion is indicated
- Uncertain diagnosis i.e. sickle cell crisis vs. osteomyelitis

Follow up

- The patient should be assessed for long-term management by a paediatrician and/or followed in a Sickle Cell Clinic.

2. Guidelines for Specific Clinical Settings:**A. Acute Painful Episode (Vaso Occlusive Disease)**

A vaso-occlusive crisis (VOC) is the most frequent complication of Sickle Cell Disease. It is a condition whereby obstruction of blood flow by sickled erythrocytes leads to hypoxia and acidosis and eventually, to ischemic tissue injury. VOC varies in intensity and duration between patients and between different episodes in the same patient. Precipitating factors may be infection, fever, acidosis, hypoxia, dehydration, sleep apnea, and exposure to extremes of heat and cold. Often, no cause is identified.

Inclusion Criteria:

- Sickle cell patients presenting with signs of VOC.
- Presenting sign relative to the site of the VOC and conditions that may mimic or occur concurrently are outlined in the table below.

Site of Pain	Signs/Symptoms	Conditions with Similar Presentations	Possible Concurrent Conditions
Bone (extremities, dactylitis, hand/foot, back)	<ul style="list-style-type: none"> • Pain • swelling • low-grade fever • redness • warmth 	<ul style="list-style-type: none"> • osteomyelitis • fracture 	<ul style="list-style-type: none"> • CNS event (stroke) • priapism • aplastic crisis • fever/sepsis • acute chest syndrome
Abdomen	<ul style="list-style-type: none"> • mild to severe abdominal pain 	<ul style="list-style-type: none"> • splenic sequestration • liver sequestration • appendicitis • cholecystitis • urinary tract infection • pelvic inflammatory disease • pneumonia (acute chest crisis) 	<ul style="list-style-type: none"> • acute sequestration crisis

Assessment and Treatment:

Emergency Management:

1. Document history and physical.
2. Monitor vital signs, oxygen saturation, clinical state and response to treatment based on severity of clinical condition.
3. Conduct the following investigations:
 - CBC, reticulocyte count
 - Lytes, urea, creatinine
 - Group and Reserve (if in severe pain, Hgb 15 g/L less than baseline, evidence of bone marrow suppression)
 - AST, ALT, gamma GT, ALP, bilirubin total and direct +/- amylase (if abdominal pain and clinical indications)
 - Arterial blood gases (if signs of respiratory failure are present)
 - Urinalysis (if indicated), culture and sensitivity if febrile
 - Chest Xray (if chest pain, fever or respiratory symptoms)
4. Encourage the patient to drink, monitor intake and output.
5. Initiate an IV (if febrile, dehydrated or in moderate to severe pain)
6. For **mild to moderate pain** give:
 - Acetaminophen (15 mg/kg/dose, maximum dose 65 mg/kg/day,) with codeine (1 mg/kg/dose, maximum dose 60 mg/dose and 6 mg/kg/day) po q4h prn. These can be given separately or together.
7. For **moderate to severe pain** (or if pain relief as given above for mild to moderate pain is inadequate after 30-60 minutes), give an IV bolus of:
 - Morphine 0.1-0.15 mg/kg/dose (maximum 7.5 mg/dose).
 - Repeat morphine once after 60 minutes if pain relief inadequate.
 - Administer a 10 mL/kg bolus of normal saline IV, followed by 1.5 times the maintenance fluid requirement of D5W with 0.45 NaCl IV.

8. For severe pain:

- Refer to Physician's Order Paediatric Continue Morphine Infusion
9. Additional IV boluses of morphine 0.05 mg/kg can be given q 1-2 h prn.
10. If adequate pain relief is established for 2 hours with 1 or 2 doses of intermittent IV morphine, consider administering acetaminophen with codeine po, as above.
11. If more than 2 doses of IV morphine are necessary, the child should be admitted to hospital.

Outpatient Management:

The child may be discharged from the emergency department if the following criteria are met:

- Taking and tolerating fluids and medication by mouth
- Pain is controlled adequately with po medications
- Concurrent problems are resolved

Children should be discharged on oral analgesics:

- Acetaminophen (15 mg/kg/dose) with Codeine (1 mg/kg/dose) po q4h prn for a period of 48 hours (acetaminophen maximum 65 mg/kg/day, and codeine maximum 60 mg/dose, 6 mg/kg/day).
- If pain persists after 48 hours, patients should be re-evaluated.

Inpatient Management:**1. Pain Control**

- Refer to Physician's Order Paediatric Continuous Morphine Infusion
- Long acting oral morphine (equivalent dose) may be used as an alternative to continuous IV morphine in stable patients
- If the child is comfortable and has graduated to demand dosing only, switch to oral analgesics: Acetaminophen (10-15 mg/kg/dose) with codeine (1 mg/kg/dose) po q4h prn (acetaminophen maximum 65 mg/kg/day, and codeine maximum 60 mg/dose and 6 mg/kg/day) given separately or together
- For unmanageable pain and/or complications consult a tertiary center and consider transfer

2. Monitoring

Children receiving IV morphine must be carefully monitored for signs of opioid toxicity which may include: hypotension, bradycardia, drowsiness, coma, pinpoint pupils, cold clammy skin, and hypoventilation. Monitoring will be according to the **Monitoring Guidelines for Patients Receiving Intravenous Opioid Analgesia: Appendix 1**

3. Additional Inpatient Treatment Recommendations

- Stool softener (e.g. docusate sodium 5 mg/kg/day) BID while on narcotic pain medication
- Antihistamines for pruritis prn
- Hydration: Continue IV/PO fluids at 1-1.5 times the maintenance rate
- Initiate oxygen protocol to treat hypoxemia (oxygen saturation \leq 94%)
- Incentive spirometry for children able to cooperate (usually older than 4 yrs of age): 10 breaths q1-2h when awake, or 5 breaths every 15 minutes

- Monitor VS q4h, fluid intake and output, and daily weight
- Monitor pain using the developmentally appropriate pain scale q4h, and before and after each pain medication and non-pharmacologic intervention (heating pads, warm baths, other comfort measures such as imagery and distraction)
- Encourage ambulation and activity as tolerated

4. Discharge Criteria

The child may be discharged if the following criteria are met:

- Taking and tolerating fluids and medication by mouth
- Pain is controlled adequately with po medications
- Concurrent problems are resolved
- Follow up with family physician or paediatrician is arranged

B. Fever (Rule Out Infection)

By 3-4 months of age (when fetal hemoglobin declines to < 50% of total), many children with sickle cell anemia (HbSS) and sickle β -thalassemia develop clinically significant hemolytic anemia and impairment of splenic function. In others, although the Hb F may remain above 50% these children are still at risk of splenic hypofunction. Even though the spleen may be enlarged during the first years of life, its phagocytic function is markedly reduced. Therefore, children with sickle cell anemia are at risk of overwhelming septicemia, often without a primary focus, due to encapsulated organisms, including *Streptococcus pneumoniae* and *Haemophilus influenzae* type B. If special measures are not taken, 15-20% of infants and young children with sickle cell anemia die before the age of 5, usually of septicemia or meningitis.

In children under 6 years of age with sickle cell disease, the predominant pathogen is *Streptococcus pneumoniae* (in 66%). In children over 6, gram-negative organisms account for over 50% of bacteremias. The incidence of pneumococcal bacteremia in children under 3 with sickle cell disease is 6 events per 100 patient-years. Furthermore, pneumococcal bacteremia carries a case fatality rate of about 20-25%.

Patients should be assessed for the following long-term management measures. Despite these, septicemia may still occur.

- Early diagnosis and referral to a comprehensive care program for sickle cell disease
- Prophylactic penicillin continued until at least 5 years of age
- If allergy (to penicillin) exists, trimethoprim-sulfamethoxazole may be used
- Immunization with the polyvalent (23-valent) pneumococcal vaccine (0.5 ml SC or IM) and quadrivalent meningococcal vaccine (0.5 ml SC) at 24 months of age
- Immunization with pneumococcal conjugate vaccine at 2, 4, 6 and 14 months of age
- Verify that routine childhood immunizations are up to date

Inclusion Criteria:

Children with sickle cell disease and a fever as defined by an oral temperature of ≥ 38 °C.

Assessment and Treatment:***Emergency Management:***

1. Place the child in a room immediately for assessment by a physician
2. Document history and physical
3. Monitor vital signs, oxygen saturation, clinical state and response to treatment based on severity of clinical condition
4. Conduct the following investigations:
 - CBC, reticulocyte count
 - Blood culture
5. **Initiate an IV and administer IV antibiotics STAT.**
Refer to **Table I: Antimicrobial Therapy Guidelines** for antibiotic selection.
6. **STAT** dose should be given within 30 minutes of presentation and before test results are available. **Do not wait for chest x-ray or blood count results to administer antibiotics.** IM injection may be used if IV access is unavailable. Parenteral antibiotics should be given even if there is an obvious focus of infection e.g. otitis media, URTI, etc.
7. Clindamycin should not be used alone in the treatment of suspected meningitis, as it does not cross the blood/brain barrier.
8. If the child is seriously ill, add vancomycin see empiric therapy for dosing guidelines.
9. Continue monitoring vital signs, capillary refill and oxygen saturation via pulse oximeter.
10. Give acetaminophen (15 mg/kg/dose, q4h, prn, maximum 65 mg/kg/day).
11. Additional investigations may be indicated only if there is clinical suspicion of other physical findings:
 - Chest x-ray, if cough, hypoxemia, tachypnea, chest pain, or toxic appearance are present
 - Arterial blood gases
 - Urine, throat or stool culture
 - Lumbar puncture
 - Group and Reserve if pale, respiratory or neurological symptoms, or splenic enlargement are present
 - Mycoplasma PCR from throat swab and Mycoplasma serology
 - Evaluation for osteomyelitis

Admission Criteria:

Hospitalization is indicated when the patient appears unwell, particularly in the presence of systemic toxicity, cardiovascular instability, **and/or** the following:

- Toxic appearance
- Recent doses of prophylactic penicillin have been missed
- Less than 1 year of age
- Respiratory distress
- Segmental/lobar infiltrate on chest x-ray
- WBC is > 30 or $< 5 \times 10^9/L$, Hgb < 60 g/L, or platelets $< 150 \times 10^9/L$
- Follow-up is uncertain (distance, inconvenience, poor compliance) or the family's ability to cope is uncertain
- Previous episodes of severe sepsis or meningitis

Table I: Antimicrobial Therapy Guidelines**STAT dose within 30 minutes of presentation and before test results are available.**

Ceftriaxone IV (80 mg/kg/dose, maximum 2 g/dose)

If allergy exists to beta-lactam antibiotics IV clindamycin (40 mg/kg/day, divided q6-8h, maximum 2.7 g/day) can be used. Should not be used alone in the treatment of suspected meningitis.

Empiric Therapy (when meningitis is not suspected)

1. IV cefotaxime (200 mg/kg/day, divided q6-8h, maximum 8 g/day) until cultures are sterile and clinical status improves (minimum of 48 h).
2. For patients with a significant beta-lactam allergy, administer IV clindamycin (40 mg/kg/day, divided q6-8h; maximum 2.7 g/day). Clindamycin is not to be used in the treatment of meningitis, as it does not cross the blood-brain barrier.
3. For children 5 years of age or older with respiratory symptoms, administer erythromycin lactobionate IV (40 mg/kg/day divided q6h, maximum 4 g/day) **OR** clarithromycin po (30 mg/kg/day divided q12h, maximum 1 g/day).
4. For children younger than 5 years of age, IV erythromycin lactobionate may be given if high suspicion of Mycoplasma.

Empiric Therapy for Presumed Pneumococcal Meningitis

1. IV ceftriaxone (80 mg/kg/dose, q12h x 3 doses, then q24h, maximum 2 g/dose) **AND** IV vancomycin (60 mg/kg/day divided q6h, maximum 4 g/day)
2. For patients with a significant beta-lactam allergy, administer IV vancomycin (60mg/kg/day, divided q6h, maximum 4 g/day) **AND** po rifampin (20 mg/kg/day, divided q12h, maximum 1.2 g/day).

Additional Treatment when Child is Critically Ill

1. IV vancomycin (60 mg/kg/day, divided q6h; maximum 4 g/day) is indicated for patients who are severely ill (septic), in whom meningitis is suspected, or who deteriorate on cefotaxime/ceftriaxone.
2. Decisions to stop vancomycin should be made in consultation with a Tertiary Sickle Cell Team.

Inpatient Management:

- Antibiotics may be changed, once culture and sensitivity results are available
- If the microbiology laboratory reports the 48 hour cultures as negative, stop antibiotics unless there is a focal infection
- If the culture is positive and the organism is penicillin-susceptible, change to IV penicillin (250,000 units/kg/day, divided q4-6h, maximum 20 x 10⁶ units/day)
- If the culture is positive for penicillin-non-susceptible pneumococcus, ensure that the patient is on vancomycin, in addition to ceftriaxone/cefotaxime and consult a Tertiary Sickle Cell Team
- If the patient has penicillin/cephalosporin-resistant pneumococcal meningitis, or is not improving after 36-48 h of therapy, do a repeat lumbar puncture as an in vivo measure of treatment effectiveness

1. Monitoring

- Observe patients closely for any deterioration in clinical status, which may indicate septicemia or development of chest crisis
- Monitor vital signs q4h
- If fever persist perform blood cultures after 24 hours and CBC daily
- Monitor reticulocyte counts every other day

2. Additional Inpatient Treatment Recommendations

- Initiate oxygen protocol to treat hypoxemia (oxygen saturation \leq 94%)
- Hydration: Administer D5W with 0.45 NaCl IV at 1.5 times maintenance for the first day (and while patients are febrile); then reduce to maintenance levels (PO+IV). Encourage oral intake.
- Administer antipyretics as necessary.

3. Discharge Criteria

The child may be discharged if the following criteria are met:

- Taking and tolerating fluids and medications by mouth
- Afebrile for at least 24 hours, with negative cultures at 48 hours
- Pulmonary symptoms, if any, have resolved
- Follow up with family physician or paediatrician is arranged

Outpatient Management:

Outpatient management of sickle cell patients with fever is an area that is still being evaluated. The Child Health Network recommends an outpatient approach in a select group of well appearing children with fever. When considering outpatient management consult the Hospital for Sick Children (HSC) Sickle Cell Team or Haematology Fellow.

Inclusion Criteria

All four criteria must be met prior to considering outpatient management

1. Patient should be assessed and shown to be free of signs/symptoms of systemic toxicity other than fever
2. Patients should receive a broad spectrum, long acting parenteral antibiotic (ceftriaxone 80 mg/kg/dose, maximum 2 g/dose)
3. The patient and family verbalize compliance and understanding of the proposed treatment plan
4. Follow up can be ensured

Once the decision has been made for outpatient management, a short period of observation in the emergency department (2-4 hours) is recommended, followed by re-evaluation prior to discharge. The following criteria should be met when outpatient management is considered:

- Recent doses of prophylactic penicillin have not been missed
- Age greater than 1 year

- WBC count between 5 and 20 x 10⁹/L; platelets > 100 x 10⁹/L
- No signs or symptoms of systemic toxicity or other sickle cell complications
- Vital signs stable
- Tolerating oral fluids
- No respiratory distress
- A dose of Ceftriaxone IV or IM has been administered.
- A verified phone number will be documented in the health record to confirm the patient's location should the health care team need to contact them.

Assessment and Treatment:

Outpatient Antimicrobial Therapy:

Following the initial **STAT** dose of IV ceftriaxone initiate: .

1. Oral antibiotics

OR

2. Return within 24 hours for a second dose of IV ceftriaxone.

Each of these approaches has its advantages and disadvantages; both are clearly dependent on patient compliance.

The patient should be given a prescription for a 3-day supply of oral antibiotic. Duration of treatment depends on the findings at reassessment, including the focus of infection.

1. Preferred Agents: *
 - cefixime (Suprax®: 8 mg/kg/day, once daily, max. 400 mg/day)
 - cefaclor (Ceclor®: 40 mg/kg/day, divided TID; max. 1.5 g/day)
2. Alternative Agents:
 - cefprozil (Cefzil®)
 - patients 6 months to 12 years of age: 30 mg/kg/day, divided BID, max. 1 g/day
 - patients >12 years of age: 250-500 mg BID
 - cefuroxime axetil (Ceftin®) 250 mg BID in tablet form (tablets and suspension are not bioequivalent and suspension is very bitter)
 - clarithromycin (Biaxin®) 30 mg/kg/day, divided BID; max. 1 g/day
 - clindamycin 30 mg/kg/day, divided q6-8h (max. 2 g/day)
3. Patients with significant allergy to beta-lactam antibiotics may be treated with clarithromycin or clindamycin

** Other antibiotics used in other centres include amoxicillin and erythromycin-sulfamethoxazole. In making the recommendations above the Child Health Network have weighed the risk of antibiotic resistance due to prior penicillin prophylaxis, the possibility of Haemophilus influenzae type B infection, and the incremental benefits of these agents in our setting in patients who have broken through penicillin prophylaxis.*

Monitoring:

The attending paediatrician will ensure that the patient is reassessed the next day through one of the following processes:

- Primary care physicians office visit
- Emergency Department visit or appointment in Paediatric Clinic
- HSC Sickle Cell Clinic visit, if recommended by the HSC Sickle Cell Team

The attending paediatrician discharging the patient will communicate to the follow up paediatrician:

- The reason for visiting the emergency department and the discharge diagnosis
- The current clinical status
- The recommended plan of care

The patient/parent will receive the discharge instruction sheet titled “Instruction Sheet for Children with Sickle Cell Disease Discharged from the Emergency Department”.

The follow up paediatrician will arrange a follow-up appointment on day 3 at the primary care physician office or HSC Sickle Cell Clinic to ensure the assessment of the following:

- Compliance with medications
- Response to treatment
- Culture results
- Duration of antibiotic therapy
- Follow up regarding penicillin prophylaxis, this should be resumed once the antibiotic therapy is completed.

Note: *Caution is recommended in managing sickle cell patients with fever as outpatients. It is suggested that only a small fraction of these patients are potentially suitable for this form of management. The strategy suggested by the Child Health Network does not represent an exclusive course of action; it will be subjected to re-evaluation and prospective evaluation.*

C. Acute Chest Syndrome or Pneumonia Management

Acute chest syndrome (ACS) is responsible for up to 25% of all deaths in children with sickle cell disease, and is the second most common cause of hospitalization in these children. The etiology is variable and may include both infections and non infectious causes. Infections are more common in younger children. The most common causes of acute chest syndrome are listed below:

Infectious Causes of ACS	Non-infectious Causes of ACS
<p>Bacteria</p> <ul style="list-style-type: none"> • Pneumococcus • Gram-negative bacteria • Chlamydia pneumoniae • Mycoplasma pneumoniae <p>Viruses</p> <ul style="list-style-type: none"> • Respiratory syncytial virus • Para-influenza • Influenza 	<ul style="list-style-type: none"> • Pulmonary infarction (in situ sickling) • Hypoventilation secondary to rib/sternal infarction or narcotic administration • Fat embolism • Pulmonary edema secondary to fluid overload

In patients with sickle cell disease, ACS occurs most frequently in patients with hemoglobin genotype SS (12.8 events/100 patient-years); less so in those with HbS β^0 -thalassemia (9.4

events/100 patient-years) or HbSC (5.2 events/100 patient-years); and least often in those with HbSβ⁺-thalassemia (3.9 events/100 patient-years) (Castro et al. 1994). Within each Hb type, the incidence is strongly but inversely related to age, being highest in children 2-4 years old (25.3 events/100 patient-years) and decreasing to its lowest value in adults.

Inclusion Criteria:

Acute chest syndrome (ACS) can be defined as a new infiltrate on chest x-ray associated with one or more new symptoms. The symptom complex may be varied, and not all symptoms are present in every episode; however, some combination of these symptoms is required for this "diagnosis." Patients with the following should be considered for ACS.

Age	Clinical and Laboratory Signs *
< 4 years	<ul style="list-style-type: none"> • Fever • Cough • Upper lobe disease
Older children and adults	<ul style="list-style-type: none"> • Shortness of breath • Chills • Severe pain • No fever • Multi lobe and lower lobe disease
All ages *	<ul style="list-style-type: none"> • Tenderness over ribs or sternum • Infiltrates in one or more lobes on x-ray (66% one lobe only)/ or chest x-ray may look normal in first 2-3 days • Pleural effusion in 30% • Decreased Hgb • Increased leukocytes

* Seasonal Variation: Increased incidence in the winter months

Assessment and Treatment:

Emergency Management:

1. If fever $\geq 38^{\circ}\text{C}$ or if in respiratory distress place the child in a room immediately for assessment by a physician. **[see guideline section B. Fever (Rule out infection)]**
2. If no fever and no breathing difficulties, assess as soon as possible (within 15 minutes, P-CTAS Level 2)
3. Document history and physical
4. Monitor vital signs, oxygen saturation, clinical state and response to treatment based on severity of clinical condition.
5. Conduct the following investigations:
 - CBC, reticulocyte count
 - Blood culture, if the child is febrile
 - Crossmatch (for possible exchange transfusion), if in respiratory distress
 - Nasopharyngeal swab
 - Chest Xray if fever, chest pain, tachypnea, or respiratory symptoms are present
6. Initiate IV Normal Saline at maintenance flow rate
7. Initiate oxygen therapy to maintain oxygen saturation $\geq 94\%$.

8. Administer IV cefotaxime 200 mg/kg/day, divided q6-8h; maximum dose 8 g/day. (may follow after initial dose of ceftriaxone in the emergency department)

Inpatient Management:

Note: All patients with ACS should be admitted to the paediatric inpatient unit at a Regional Children’s Health Centre or transferred to tertiary care center depending on the severity of the child’s condition.

1. Antimicrobial Therapy:

- For the first 72 hours of admission, administer IV cefotaxime, 200 mg/kg/day, divided q6-8h, maximum 8 g/day starting after x-ray results reviewed, may follow initial admission dose of ceftriaxone
- Beyond 72 h, some may be switched to IV cefuroxime (75 mg/kg/day, divided q8h, maximum 4.5 g/day), as follows:

Mild pneumonia and stable	Cefotaxime for 72 h, then cefuroxime
Moderately severe pneumonia	Continue cefotaxime
Severe pneumonia or unstable	Transfer to a tertiary center. Continue cefotaxime and add vancomycin 60 mg/kg/day, divided q6h, maximum 4 g/day

- Children \geq 5 years of age should be suspected of having mycoplasma pneumonia; add erythromycin lactobionate IV (40 mg/kg/day divided q6h, maximum 4 g/day) **OR** clarithromycin po (30 mg/kg/day divided q12h, maximum 1 g/day). Use IV erythromycin in patients younger than 5 only if there is evidence of mycoplasma
- For patients with a significant beta-lactam allergy, IV clindamycin (40 mg/kg/day, divided q6-8h; maximum 2.7 g/day) or po clindamycin (30 mg/kg/day, divided q6-8h; maximum 2 g/day) can be administered.

2. Transfusion Guidelines:

For all patients being considered for transfusion, the Paediatrician will contact the Hematology consult fellow (or, after hours, the Hematology/Oncology fellow on call) to discuss treatment options and the need for transfer to a tertiary center.

- Patients with **mild to moderate disease** and hemoglobin (Hgb) at baseline **do not** generally need a transfusion.
- Patients with **moderately severe disease** and Hgb 15 g/L less than baseline should be transfused with packed red blood cells, 10 mL/kg (simple transfusion).
- Patients should not be transfused to a Hgb of greater than 100 g/L (Hct > 30%)
- Patients with **severe disease**-extensive infiltrates, worsening ABGs, increasing need for oxygen (> 40% O₂) and decreasing O₂ saturation should have an **exchange** transfusion (RBC cytapheresis). **Arrange transfer immediately to a tertiary care center.** Patient may require the initiation of transfusion to stabilize.

3. Monitoring:

- Monitor vital signs
- Continuous oxygen saturation monitoring if the patient is on oxygen therapy
- Observe patients closely for any deterioration in clinical status, which may necessitate transfer to a tertiary center.

- Daily CBC and arterial/venous blood gases (ABG/VBG) if unstable. If not improving, consider transfusion according to the **transfusion guidelines**.

4. Additional inpatient treatment recommendations:

- Provide analgesic according to the guidelines in section A: Acute painful episode (vaso occlusive disease) management.
- Administer antipyretics as necessary.
- Continue IV and PO fluids at maintenance flow rates. Increase fluids as needed, if the child is dehydrated or insensible losses are increased (e.g., persistent fever).
Caution: Excessive fluids may precipitate or exacerbate ACS.
- If clinical or radiographic signs of fluid overload are present, administer IV furosemide 0.5 -1 mg/kg/dose, maximum 60 mg/dose.
- If the child has a history of reactive airway disease or wheezing, consider bronchodilators, e.g. salbutamol.
- Incentive spirometry for children able to cooperate (usually older than 4 yrs of age): 10 breaths q1-2h when awake, or 5 breaths every 15 minutes.
- Encourage ambulation and activity. Involve the Child Life Worker as necessary to recommend structured daily activity.

5. Discharge Criteria:

The child may be discharged if the following criteria are met:

- The child does not require supplemental oxygen
- The patient has been afebrile for at least 24 hours
- The child is taking fluids and medications by mouth
- Pain control is adequate with PO medications
- Concurrent problems are resolved
- Follow up with a family physician or paediatrician is arranged

Appendix 1. Monitoring Guidelines for Patients Receiving Intravenous Opioid Analgesia:

Analgesia	Monitoring			
	Observation/Vital Signs/ Pain Score/Sedation Scale	Oxygen Saturation	Resp/ Apnea	ECG
Continuous Opioid Infusion	<p>Baseline Assessment:</p> <ul style="list-style-type: none"> ▪ HR, BP, RR, sedation score and pain score <p>Initiation Assessment:</p> <ul style="list-style-type: none"> ▪ HR, BP, RR, sedation score and pain score q30min. x 2 then q1h x 2. <p>Then:</p> <ul style="list-style-type: none"> ▪ RR q1h ▪ HR, BP q4h, pain score q4h while awake and prn ▪ Sedation score q8h and prn ▪ After change in rate or bolus, repeat initiation assessment 	Continuous monitoring	As ordered	No
Intermittent Opioid Dose	<p>Baseline Assessment:</p> <ul style="list-style-type: none"> ▪ HR, BP, RR, O2 Sat, sedation score and pain score <p>Administration Assessment: (<i>usual infusion rate per dose, 15-20 minutes</i>)</p> <ul style="list-style-type: none"> ▪ HR, BP, RR O2 Sat and sedation score q 15 minutes x 2 then q 30 minutes x 2 ▪ Pain score at the end of the flush <p>Then:</p> <ul style="list-style-type: none"> • as per patient condition/other pre-existing orders 	Continuous monitoring for patients whose underlying condition predisposes them to respiratory depression	As ordered	No

Sedation Score:

0 Awake

1 Mild – arousable with verbal stimulation

2 Moderate – arousable with tactile stimulation

3 Severe – not arousable by tactile stimulation

Evaluation:

- Retrospective case review to assess the outcome for children with fever managed as an outpatient
- High risk case review process
- Evaluate compliance with the use of the pre printed physician order Paediatric Sickle Cell Disease

References:

1. Child Health Network: Management of Sickle Cell Disease in Children. Practice Guideline 2004-2004.
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