CancerCare Manitoba
ActionCancerManitoba

CancerCare Manitoba Clinical Practice Guidelines

Provincial Consensus on the Urgent Management of Acute Complications of Sickle Cell Disease in Pediatric Patients

Version 1.0 – July 2025

CancerCare Manitoba Guideline
Disease Management – Sickle Cell Disease, Acute Complications, Pediatrics
Developed by: Outpatient Clinic Services Program and Pediatric Hematology

1.0 Background

Aim and Purpose

Development of this clinical guide was undertaken for the purpose of knowledge translation of the current standards in practice for the identification of and interventions for acute complications of sickle cell disease in pediatric patients. The overall aim is to improve the standard of care received by this patient population, through application of standardized evidence-based interventions and promotion of best practices.

Development Process

Local practices and international consensus guidelines on identification and management of acute complications of sick cell disease in pediatric patients were reviewed. Based on the evidence evaluated, a guideline was drafted and presented to CancerCare Manitoba (CCMB) Outpatient Clinic Services and Systemic Therapy Programs and pediatric hematologists for review. The following consensus guideline was produced following internal review.

Patient Population and Healthcare Setting

The recommendations in this clinical guide are applicable to pediatric patients who are experiencing acute complications of sickle cell disease.

For this purpose, it may be used by qualified and licensed healthcare practitioners involved with the care of pediatric patients, which may include (but is not limited to): CancerCare Manitoba physicians, surgeons, nurses, pharmacists at CCMB, as well as health care providers in the community and at Health Sciences (HSC) Winnipeg Children's Hospital. It should be noted that most children in Manitoba experiencing acute complications of sickle cell disease will not be managed at CCMB. They are more likely to be managed in the community, in regional emergency departments or at HSC Winnipeg Children's Hospital.

Disclaimer

This guideline document should be viewed as an evidence-informed practice tool, and as such, it does not represent an exhaustive text on acute complications of sickle cell disease. Clinicians are advised to use it in their practice concomitantly with information from other evidence-informed sources.

Use of this guideline in the clinical setting should not preclude the use of the practitioner's independent clinical judgment. It is the responsibility of the practitioner to develop an individualized disease or symptom management plan for each patient under their care, and ideally, this should take place within the context of a multidisciplinary team. The needs and preferences of the patient and the family should always be reflected in the plan of care.

2.0 Methodology

Working Group

- Working Group Members: Dr. Sara Israels, Lucas Tielman, Trisha Vera, Cathy Bourne, Dr. Ashley Chopek, Dr. Kyle Mendonça, Tori Scott, Theresa Whiteside, Chris Vargek
- Reviewers: Tracey Schlamb, Katrin Ann Param-Vital, Dr. Chantalle Menard, Dr. Leonard Minuk
- Conflicts of Interest: None

Literature Search

Recommendations were adopted from the Translating Emergency Knowledge for Kids (TREKK.ca). Guidance from the Canadian Haemoglobinopathy Association, The Hospital for Sick Children, and local practices in Manitoba were also considered. After review and revisions, a consensus was reached among CCMB staff involved in identifying acute complications of sickle cell disease in pediatric patients and recommended nursing interventions.

Maintenance

At CancerCare Manitoba clinical guidelines are considered 'living' documents which require ongoing evaluation, review and updating. Re-evaluation of this clinical guide is planned for 2027. The working group will revise and update the document as needed, with any critical new evidence brought forward before this scheduled review.

3.0 Definitions & Management Algorithms

Types of acute complications of sickle cell disease (SCD)

- Fever
- Vaso-occlusive episode (VOE)
- Acute chest syndrome (ACS)
- Aplastic anemia
- Stroke
- Splenic sequestration
- Priapism

Emergent Recommendations

1. Febrile patients should have antibiotics initiated within 30-60 minutes of arrival.

   See *Table 1* for antibiotic recommendations.

2. Pain management should be initiated within 30-60 minutes of arrival.

   See *Table 2* for analgesic recommendations.

Fever (Temperature ≥ 38ºC)[2]

Infection is the most common cause of SCD-related mortality.

- Febrile patients require prompt investigation and intervention.
- Children with SCD require extended immunizations because of functional asplenia. If under/unimmunized, they are at an even higher risk of serious bacterial infection.
- Intravenous (IV) antibiotics should be given. A 2nd or 3rd generation cephalosporin is recommended for initial treatment. Additional antibiotics should be chosen based on the clinical picture, as outlined in *Table 1*.
- Identify signs of sepsis including poor perfusion, hypotension, hypoxemia, tachycardia and/or altered mental status and notify on-call pediatric hematologist.
- Admission for ongoing IV antibiotics and supportive care are indicated for patients meeting any of the high-risk criteria below. If patient has no high-risk criteria AND reliable follow up of blood culture results, discuss admission vs. outpatient management with Pediatrics/Pediatric Hematology/Pediatric Referral Centre. Admit to hospital if blood culture results are positive at any time.

High Risk Criteria in a Febrile Patient[2]

• Hemodynamic instability or unwell appearance • Fever >40 Degrees Celsius (°C) • Age <6 months • Respiratory distress • Leukopenia (WBC <5x10^9/L), leukocytosis (WBC >30x10^9/L), thrombocytopenia (PLT <100x10^9/L), or significant anemia (<60g/L or a ≥20g/L drop from baseline)	• Severe illness including meningitis, osteomyelitis, acute chest syndrome, splenic sequestration • Previous history of pneumococcal sepsis and/or meningitis • Severe pain that cannot be managed at home • Unable to arrange close follow-up

Vaso-Occlusive Episode (VOE)[2]

The most common presentation and frequent cause for hospital admission is pain. VOE can be triggered by dehydration, illness, fever, surgery, cold exposure, and stress.

- Pain control should be initiated as soon as possible (see *Table 2* for dosing).
- Consider splenic sequestration in all patients with abdominal pain.
- Dactylitis (see *Figure 1*), VOE of hands and feet is more commonly seen in infants.
- Consider other painful conditions in SCD such as osteomyelitis, acute vascular necrosis, acute chest syndrome, stroke, as well as non-SCD related causes of pain.
- For uncomplicated VOE, there is no requirement for investigations. Decisions regarding investigations should be made on a case-by-case basis.
- Utilize the '3 Ps' approach to pain management; pharmacological, psychological, and physical therapeutic.[1]

Figure 1. Dactylitis

Acute Chest Syndrome (ACS)[2]

ACS is a potentially life-threatening complication of SCD, and a leading cause of morbidity.

Diagnostic Criteria:

1. Fever, respiratory symptoms including cough, wheeze, dyspnea, hypoxia, or chest pain AND,
2. New pulmonary infiltrate involving >1 lung segment (not atelectasis)

Children with asthma and SCD are twice as likely to develop ACS as children with SCD alone. Symptoms of ACS overlap with lower respiratory tract infections and asthma.

Initial Management:

- Apply oxygen via mask/nasal prongs to maintain SpO2 ≥95%. Consider treating patients with a history of asthma using bronchodilators.
- **Contact Pediatric Hematology via HSC paging (204-787-2071).**
- Will require analgesia but avoid over-sedation from analgesics leading to hypoventilation.
- Do not give more than maintenance fluids as overhydration can exacerbate ACS.
- Obtain blood cultures and nasopharyngeal swab for viruses and start antibiotics.

***All patients with ACS will require hospital admission to provide critical care support, monitoring and consideration of Red Blood Cell (RBC) exchange.**

Aplastic Crisis[2]

Severe acute anemia can be caused by any viral illness, particularly parvovirus B19. Symptoms may include fever, fatigue, headache, and pallor. Severe reticulocytopenia is seen (reticulocytes <1%).

Blood transfusion may be indicated for management if anemia is severe. Management decisions including the need for RBC transfusion should be discussed with Pediatric Hematology.

Stroke[2]

Stroke is a serious complication that should be investigated for any new neurologic deficit. SCD is the most common cause of stroke in children.

- Headache, sudden weakness, abnormal speech, seizures, abnormal gait, and/or any focal neurologic deficit are red flags for stroke.

***For suspicion of stroke**, direct patient to nearest Emergency Department (ED) immediately, and contact the Pediatric Hematologist on-call.

Splenic Sequestration[2]

Splenic Sequestration is the result of splenic enlargement and a sudden drop in hemoglobin. It is caused by blood trapping in the spleen and can lead to hypovolemic shock. Children ages 6 months to 5 years are at highest risk, but it can occur in older children, particularly those with Hemoglobin SC disease.

Symptoms include:

- Weakness, pallor, hypotension, tachycardia, tachypnea, left upper quadrant pain and splenomegaly.
- Atypical symptoms such as back, left flank, and/or chest pain can occur.

Initial management:

- If the patient is hypotensive, IV fluid for volume support should be started immediately.
- RBC transfusion is likely to be required, but should be discussed with Pediatric Hematology.

***For any suspicion of splenic sequestration**, direct patient to nearest Emergency Department and contact the Pediatric Hematologist on-call.[1]

Priapism[2]

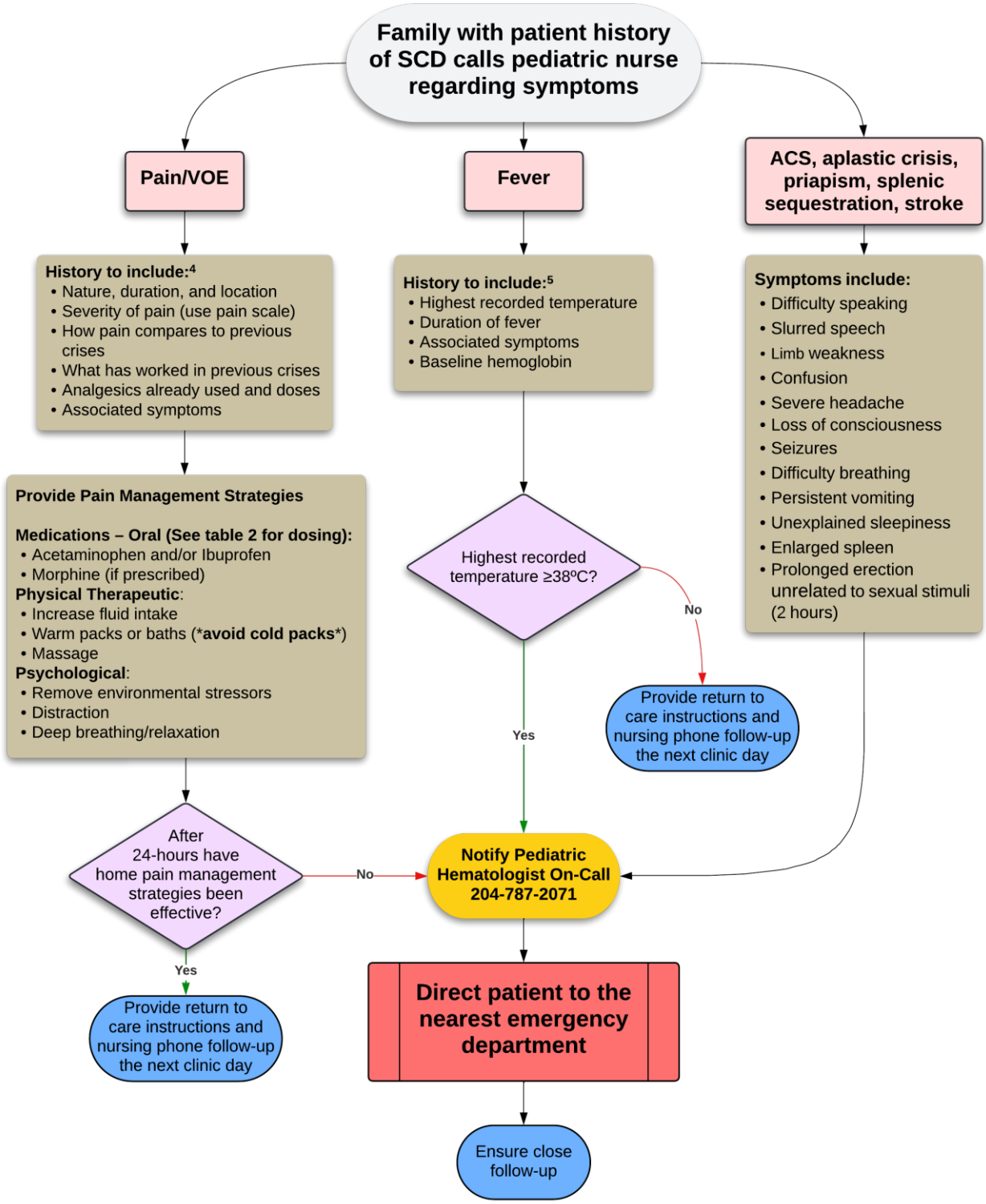
Sickle cell disease is the most common cause of priapism in children and is defined by a prolonged erection unrelated to sexual stimuli. Initial assessment includes time from onset, presence of pain, and precipitating events.

Supportive care includes IV hydration, analgesia, and supplemental oxygen to keep SpO_2 ≥95%.

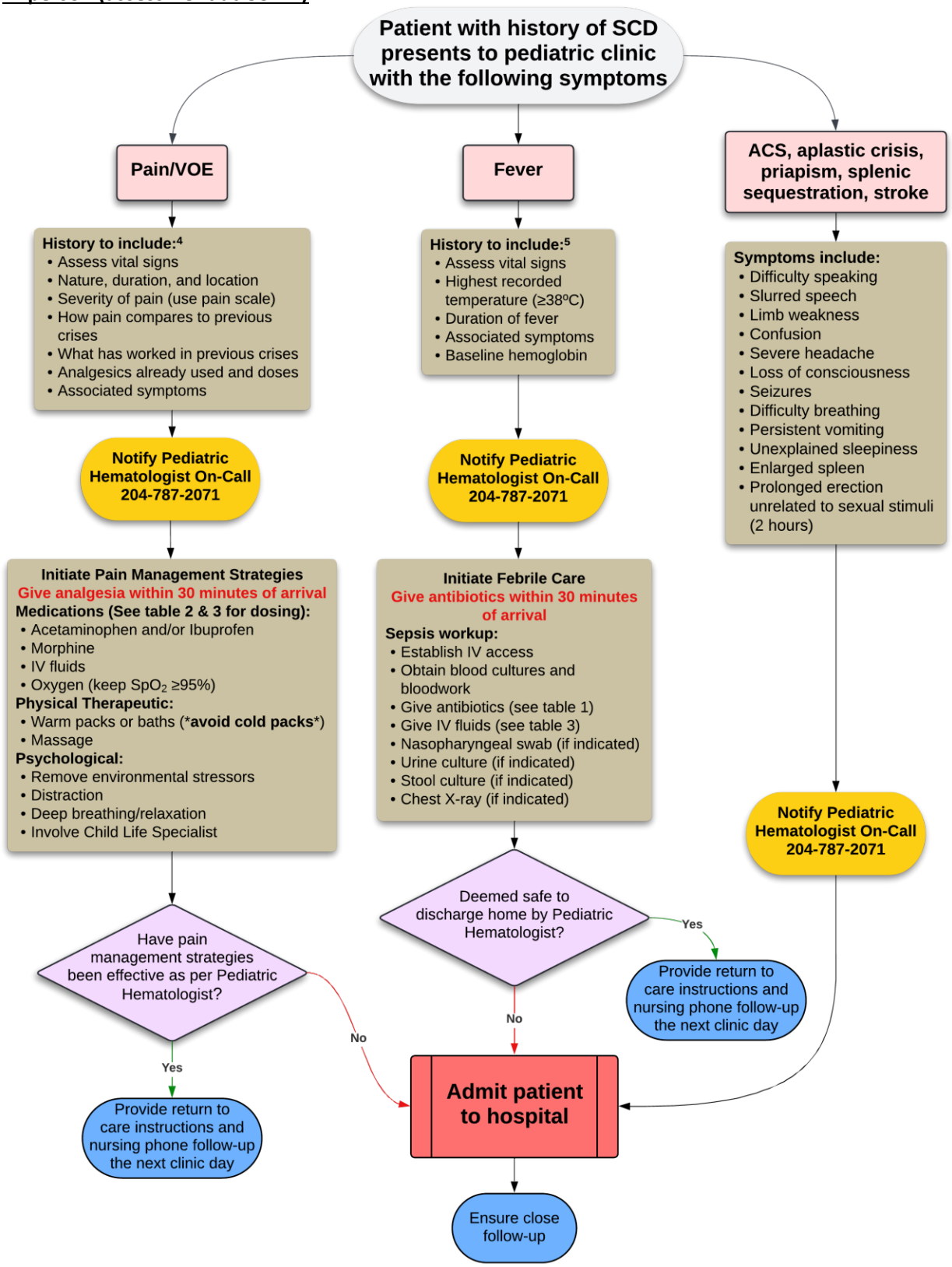
***Priapism lasting ≥4 hours is a surgical emergency**: direct patient to nearest Emergency Department immediately and contact the Pediatric Hematologist on-call. Expertise from Pediatric Urology will be required.

Symptom Evaluation and Management

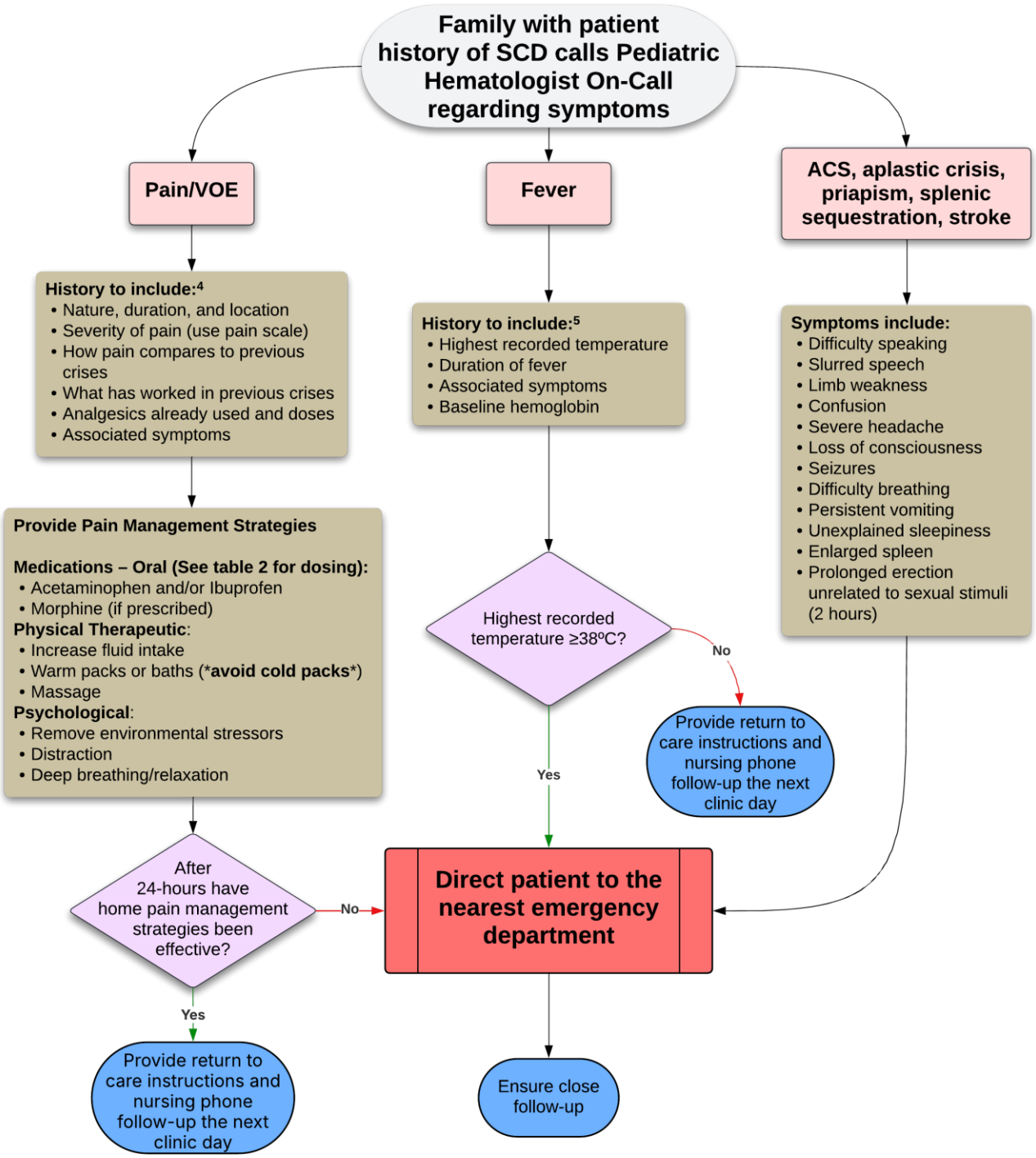
Phone Call (patient is outside of CCMB)



In-person (assessment at CCMB)



After-hours & Weekends (patient is outside of CCMB)



4.0 Related Documents

Education

For patient education materials, wallet card and standards of care for sick cell disease, please go to the CCMB SharePoint at: CCMB SharePoint ➷ Department of Pediatric Hematology & Oncology ➷ Documents ➷ Hemoglobinopathies – Standards of Care.

Tables

Table 1. Antimicrobials for Sepsis and Acute Chest Syndrome (Age >1 month)[3]

Drug		Dose	Indication
INITAL	Ceftriaxone	100 mg/kg/dose (MAX 2000 mg/dose) IV q24h *Meningitic dose:* 100 mg/kg/dose (MAX 2000 mg/dose) IV x 1 dose then 12 hours later 50 mg/kg/dose (MAX 2000 mg/dose) IV q12h	Initial antibiotic of choice for sepsis and ACS. Includes salmonellae coverage if osteomyelitis is suspected.
ALTERNATIVE	Clindamycin	40 mg/kg/**day** IV divided q8h (MAX 900 mg/dose)	Alternative to ceftriaxone if beta-lactam allergy is suspected.
ADJUNCT	Vancomycin	15 mg/kg/dose (MAX 1000 mg/dose) IV q6h	Add if Methicillin-resistant Staphylococcus aureus (MRSA) risk factors present, if clinically unwell or for patients with hemodynamic instability.
	Azithromycin	10 mg/kg/dose (MAX 500 mg/dose) on Day 1, followed by 5 mg/kg/dose (MAX 250 mg/dose) for remainder of treatment	Add for all patients > 5 years of age with respiratory symptoms. Consider in < 5 years if high suspicion for mycoplasma.
	Oseltamivir	*< 1 year:* 3 mg/kg/dose (MAX 30 mg/dose) PO BID *1 year or older:* ≤ 15 kg: 30 mg PO BID >15 – 23 kg: 45 mg PO BID > 23 – 40 kg: 60 mg PO BID > 40 kg: 75 mg PO BID	Consider addition if high suspicion for influenza.

Table 2. Analgesic medication dosing recommendations for VOE[1]

Drug	Dose	Indication & Comments
Acetaminophen	15mg/kg/dose every 4 hours	Mild to moderate pain
Ibuprofen	10mg/kg/dose every 6 hours	Mild to moderate pain
Morphine	Oral: 0.2-0.5mg/kg/dose every 3-4 hours (MAX 15 mg/ dose) IV: 0.05-0.1 mg/kg/dose every 2-4 hours (MAX 4-8 mg/ dose)	Moderate to severe pain

Table 3. Hydration dosing recommendations[2,5]

IV Fluid	Dose	Indication & Comments
Normal saline (NS)	10-20mL/kg of 0.9% NS given over 1 hour, if there is evidence of clinical dehydration.	Avoid over hydration (increases risk for ACS). If available, give 0.45% NS instead.

Legend

BID = twice a day
KG = kilogram
MG = milligram
ML = milliliter
PO = by mouth

References

1. Ali, S., Chumpitazi, C., Drendel, A. L. & Poonai, N. (2023, June). *Bottom Line Recommendations: Pain Treatment. Version 4.0*. Trekk - Translating Emergency Knowledge for Kids.
2. Canadian Haemoglobinopathy Association. *Consensus Statement on the Care of Patients with Sickle Cell Disease in Canada*. Version 2.0. Ottawa; 2015. https://www.canhaem.org/wp-content/uploads/2018/05/Sickle-Cell-Consensus.pdf.
3. Kesselman R., Stoffman J., Nurmi M A. (2023, June). *Bottom Line Recommendations: Sickle Cell Disease. Version 1.0*. Trekk - Translating Emergency Knowledge for Kids.
4. SickKids – The Hospital for Sick Children. (2021, July 21). *Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease. Version 5*. https://wapps.sickkids.ca/clinical-practice-guidelines/clinical-practice-guidelines/Export/CLINS214/Main%20Document.pdf.
5. SickKids – The Hospital for Sick Children. (2021, July 21). *Fever: Guidelines for Management in Children with Sickle Cell Disease. Version 5*. https://wapps.sickkids.ca/clinical-practice-guidelines/clinical-practice-guidelines/Export/CLINS215/Main%20Document.pdf.
6. Umana J, Lapite A, Ellison AM. *New horizons in emergency department management of pediatric sickle cell disease.* Ped Emerg Care 2024; 40:406-1

Contact Physician

Dr. Sara Israels

Pediatric Hematologist-Oncologist ,CancerCare Manitoba

sisraels@cancercare.mb.ca

Clinical Practice Guideline Initiative

Contact
CCMBCPGI@cancercare.mb.ca

Dr. Chantalle Menard

Haematologist, CancerCare Manitoba

Tracy Thiele

Director, Quality & Patient Safety, CancerCare Manitoba

Amanda Bak
Clinical Practice Guideline and Patient Safety Coordinator, CancerCare Manitoba

Approved By

Dr. Donna Turner
Interim President and CEO, CancerCare Manitoba

Dr. Arbind Dubey
Chief Medical Officer, CancerCare Manitoba

Ken Borce

Chief of Clinical Operations, CancerCare Manitoba

CancerCare Manitoba
675 McDermot Avenue
Winnipeg, Manitoba, Canada
R3E 0V9
www.cancercare.mb.ca

CCMB Clinical Practice Guideline: Nursing
Urgent Management of Acute Complications of
Sickle Cell Disease in Pediatric Patients
Version 1.0 July 2025